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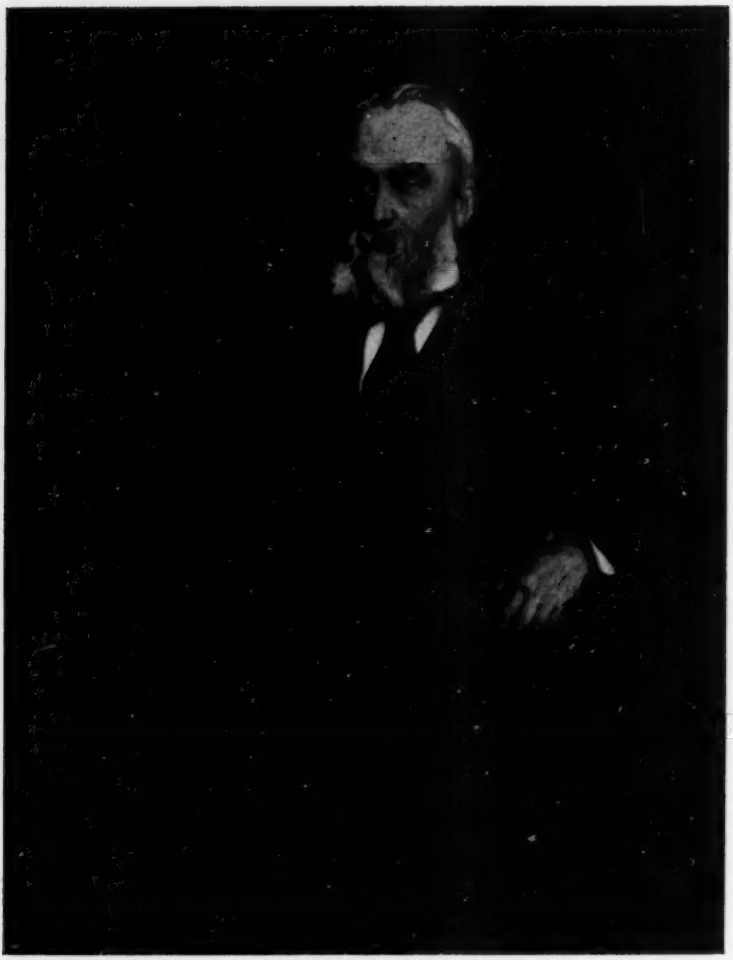

HUGHLINGS JACKSON

(From a painting at the National Hospital, Queen Square.)

Copies of this portrait can be obtained from the Editor.

HUGHLINGS JACKSON*
(1835—1911)

The major advances in knowledge are made by experimental investigation, but it is sometimes forgotten that this must be preceded by experiment in thought, and it was in this that Jackson's genius was transcendent. From clinical observation he arrived at questions that have provided the material for an immense amount of experimental research in the laboratory, as well as guiding clinical enquiry in profitable directions. To take examples, his conclusion that positive symptoms from destructive lesions must arise from the uncontrolled activity of the healthy parts remaining—simple as it now appears—was the starting point of a revolution in thought which has ever since inspired neurophysiology. Again, if we consider our present knowledge of the epilepsies, Jackson's concept of the discharging lesion and his idea that there are as many different kinds of epilepsy as there are sites of discharge were the seeds from which the trees have grown.

SIR CHARLES SYMONDS.

JOHN HUGHLINGS JACKSON was born in the village of Green Hammerton, near Knaresborough, Yorkshire, on April 4, 1835. His father, Samuel Jackson, was a yeoman farmer who also carried on a business as a brewer. After attending local schools, John was apprenticed to William Anderson, a surgeon in York, and he attended the York School of Medicine and Surgery, a small but excellent centre of instruction which played an important part in the development of provincial medical education. From there he went to St. Bartholomew's Hospital, London, where Sir James Paget was one of his teachers, and in 1856 he qualified as M.R.C.S., L.S.A. He returned to York as house-surgeon at the Dispensary and remained there for three years. In 1859 he migrated to London, armed with an introduction to Mr. (later Sir) Jonathan Hutchinson, a fellow Yorkshireman.

At this stage in his life Jackson, who had become a keen student of Herbert Spencer, thought of giving up medicine to devote himself to philosophy, but Hutchinson dissuaded him and helped him to obtain an appointment at the Metropolitan Free Hospital. Jackson's progress was now rapid. In 1862 he was elected assistant physician to the National Hospital in Queen Square, becoming full physician in 1867. He was appointed assistant physician to the London Hospital in 1863, having already lectured for some time in its medical school, and promoted to physician in 1874. He was also connected with the Islington Dispensary and with Moorfields Eye Hospital, where he was one of the first to use the ophthalmoscope in the diagnosis of brain disease. Having taken the M.D. St. Andrews and the M.R.C.P. in 1860, he was elected F.R.C.P. and F.R.S. in 1868. Some of his most important work was reported in the Goulstonian, Croonian, and Lumleian Lectures which he gave before the

College of Physicians. Jackson died at No. 3, Manchester Square, London, on October 7, 1911, as is recorded on the commemorative plaque set up by the London County Council to mark the home of this eminent Londoner.

It would be impossible to record the great neurologist's achievements in a brief note, but it may be said that his main work was concerned with speech defect in brain disease, with the epilepsies, and with the doctrine of 'levels' in the central nervous system. All his work was based on an immense amount of detailed observation and recording, and it combines attention to the minutest detail with a power of the widest generalisation. He is, by general consent, awarded the first place among those who have contributed to neurology as a science, and his stature continues to grow with the passage of time. Personally, he was a most lovable character—modest, simple and unfailingly kind—and not without some of those little eccentricities which are often found in men of genius.

W. J. BISHOP

EARLY DAYS OF THE NATIONAL HOSPITAL, QUEEN SQUARE

On November 2, 1959, a reception was held at the Hospital to commemorate the precise centenary of the inaugural meeting which led to the foundation of the National Hospital for the Relief and Cure of the Paralysed and Epileptic. At this party Sir John Woods, the hospital's chairman, asked Dr. Macdonald Critchley, who, he said, 'knows more than anybody else about the history of the hospital and its locality', to tell the guests a little about it. Dr. Critchley replied in these words:

JUST over a century ago there lived in St. Pancras, in quite modest circumstances, a small but close-knit family comprising a brother and two sisters, Edward Chandler, Johanna Chandler and Louisa Chandler. They had been left orphans at an early age and were brought up by a much beloved grandmother.

It so happened that this grandparent was taken ill and had to leave St. Pancras for a time. When she came back she was paralysed and the Chandler family realised the striking lack of facilities, medical and nursing, that existed at that time, for looking after chronic neurological patients. And they also realised that if it was a calamity for them, the plight must have been worse for those in humble and poor circumstances.

The old lady died and Louisa and Johanna were fired with a determination to rectify this gap in the social order. They spent their spare time making little ornaments of shells, beads and pearls, which they sold, and over the next two years they accumulated about £200 in this way.

By the spring of 1859 they approached the then Lord Mayor of London, David Wire, who himself had had a stroke and was partially paralysed. The Lord Mayor was sympathetic and co-operative, and he got in touch with his influential friends and business acquaintances, and interested them in the project. On November 2, 1859, he called a special meeting in the Egyptian Hall of the Mansion House, at which it was decided to found a special hospital for the investigation, care and treatment of patients suffering from paralysis and epilepsy. A small *ad hoc* committee —what we today would call a Working Party—was formed, with the Lord Mayor as chairman, Viscount Raynham as treasurer and Edward Chandler as secretary.

The committee met every week in the City and one of its first tasks was to find suitable accommodation. By the spring of 1860 they had found a house available at 24 Queen Square, which they leased at £110 a year. They invited two physicians to join the staff as honorary consultants—Dr. Jabez Ramskill and that brilliant but eccentric American-French celebrity Dr. Brown-Séquard. These formed the first medical staff and shortly afterwards they were able to enlist the sympathy of the surgeon-in-ordinary to the Queen, Sir William Ferguson, who consented to come on the staff as consulting surgeon. Patients were found and admitted in May 1860 and there is a note in the old minute book, dated March 14, 1861, to the effect that Mary

Warwick, of No. 7 Hayes Court, Soho, the first patient received in the hospital, was discharged cured, and she tendered her thanks for all the benefits received.

From the very start lectures were delivered in the Board Room by Brown-Séquard, and the hospital grew rapidly in size, importance and prestige. Gradually, when funds made it possible, they extended their premises, and within 18 months they were trying to buy Queen Square House next door, the residence of the then Lord Chief Justice, Baron Pollock, where the invalid King George III had been nursed.

They then bought a house next door, as well as the premises in the rear, in Powis Place. A little later they incorporated 26 Queen Square, a house belonging to William Morris, where his factory had been set up. Over the next few years they purchased most of the property on the east side of Queen Square, including number 32, which was part of St. Katherine's Convent for Benedictine nuns.

I would like to read you this advertisement, recorded in the minutes for March 17, 1864:

> Wanted, a Matron for The National Hospital for Paralysis and Epilepsy, Queen Square, Bloomsbury. A well-educated protestant lady of evangelical principles, aged 30 to 45, to act in above capacity. She will be required to take the entire charge and direction of the establishment, its nursing service and domestic affairs. Preference will be given to one who has already filled a similar position. Salary commencing at £50 a year, besides apartments, board, lodgings, washing, beer. No lady need apply whose character will not bear the strictest enquiry. Applications by letter only to be addressed to the Deputy Chairman, St. Paul's Churchyard.

MACDONALD CRITCHLEY

THE CONTRIBUTION OF HUGHLINGS JACKSON TO NEUROLOGY

MACDONALD CRITCHLEY, M.D.(BRISTOL), F.R.C.P.

Senior Physician to the National Hospital, Queen Square, and Senior Neurologist to King's College Hospital, London

In this country neurology took origin not from psychiatry but from general medicine, with anatomy and physiology as its deeper foundations. Jackson's work has endowed British neurology with certain national characteristics. His teaching emphasised the supreme importance of close clinical observation, coupled with a completely honest reportage of the evidence. On this basis Jackson added something peculiarly his own—a quality of vision.

Hughlings Jackson joined the staff of the National Hospital, Queen Square in 1864, when the so-called era of degradation in medicine was drawing to a close. A sort of false dawn in neurology had just broken, thanks to the morphological studies of Charles Bell, the experimentation of Marshall Hall, and the clinical flair of Robert Bentley Todd. The stage was set, but the curtain had not yet risen.

Jackson's genius was assisted by the conjugation of his immediate colleagues, who were men of outstanding ability—Gowers, Horsley, Ferrier, Brown-Séquard; and, to a lesser extent, Bastian, Beevor, Lockhart-Clarke, Broadbent and Ogle. Their professional talents differed from Jackson's, but they amplified and simplified Jackson's ideas and helped to bring some of them to fruition. Thus his views on cerebral function, which were largely the product of pure reason, were validated by Ferrier in the laboratory. In turn this work inspired the birth of neurosurgery and led directly to bold operative attacks on certain types of brain-lesion. Consequently it would be foolish to try and draw an antinomy between Jackson and Gowers, for example, for their minds were different. Their contributions to neurology were complementary rather than contrastive.

In his emphasis on the essential importance of precise observation at the bedside Jackson was not alone, for Gowers shared the same thoroughness of approach. As a house-officer Jackson had learnt the value of making post-mortem checks on his diagnoses, and he faithfully attended the autopsies of cases which had been under the care of his chiefs. Again, like Gowers, Jackson had enthusiastically accepted and utilised the new diagnostic technique of ophthalmoscopy. Both were able draughtsmen, and each made and preserved his own atlas of fundus appearances.

Besides this keen, careful and unhurried observation at the bedside, Jackson laid stress on the necessity for recording in full, without embellishment or pseudo-interpretations. The observer, he said, must set down in his notes exactly what it was the patient did and said, and not a rationalisation thereof. For example, if a patient did not protrude his tongue when told to do so, the simple fact should be stated; it would be wrong to speak of a 'paralysis of the tongue'. This rigorous discipline in observation and note-taking was urged again and again in Jackson's writings. It was this same honesty in clinical methods which inspired Jackson with a number of novel ideas which would have been smothered had he not followed his own clinical precepts.

Jackson was influenced by the revolu-

tionary ideas in science which were in the air during his student days. The biological theories of Russell Wallace and Darwin had been seized on by Jackson's mentor in philosophy—Herbert Spencer. Spencer's hypotheses led to Jackson's conception of a process of evolution going on within the nervous system. This acted in reverse during cerebral disease, and according to the intensity of the morbid process, so one faculty after another might be shed like the leaves of an artichoke. Thus emerged his idea of dissolution within the nervous system. The phenomena of, say, inebriety, would produce a succession of events recapitulating the building-up of nervous function in the growing individual. Here was a somewhat unorthodox manner of interpreting neurological symptoms, for Baillarger's work along the same lines was not generally known. The Baillarger-Jackson conception has since become insensibly incorporated in established neurological thinking.

Bound up with the doctrine of evolution and dissolution of the nervous system arose the idea of levels of function. Jackson recognised three such levels but did not identify them clearly enough to avoid frequent misunderstandings. The levels were complex, and not wholly anatomical or physiological entities. Thus, in his hierarchy within the nervous system, we find: (1) a lowest level, which consisted of the spinal cornua, Clarke's column, Stilling's nucleus, and the homologues of these parts; (2) a middle level, consisting in the motor and sensory regions of the brain as well as the corpus striatum; and (3) the highest level, made up of the prefrontal and occipital lobes, together forming the 'organ of mind' or the physical basis of consciousness. Just as dissolution might skim off one layer after another, so also epilepsy—which to Jackson entailed a 'discharging lesion'—might arise at any one of these neuronic levels.

Jackson was one of the first to distinguish between positive and negative manifestations in neurological disease. Positive clinical phenomena—e.g. an involuntary movement—could not proceed directly from negative lesions of the brain. A negative lesion, however, might *permit* a positive symptom to appear through the removal of some normal inhibitory process of restraint. For Jackson all symptomatic conditions were duplex. Thus a patient with aphasia due to cerebral thrombosis might utter wrong words. That the softening caused the defect of speech was, according to Jackson, admissible; but an area of softening could not possibly cause wrong utterances: these must surely emanate from the activity of intact parts of the brain.

Another of his *obiter dicta* was to point out the fallacy of identifying the cerebral localisation of a symptom-producing lesion with the 'localisation' of a normal function. This was a common error in thinking throughout the nineteenth century, and even later. A notable instance lay in the domain of aphasia. Jackson admitted that a lesion of Broca's region could bring about a disorder in speaking; but to argue from this that Broca's region constituted a 'centre' for articulate speech was wholly unjustifiable.

There was yet another of his ideas that attracted attention only after his lifetime. This was the notion of the 'four factors in the insanities'. Using 'insanity' in an unfamiliar fashion to connote almost any symptom engendered by brain-disease, Jackson pointed out that the clinical picture was a resultant of at least four aetiological variables, namely: (1) the depth of the dissolution; (2) its rapidity; (3) the influence of internal and external factors on the patient; and (4) the kind of brain in which the dissolution occurs—whether the patient was a child, an adult or an old man; clever or stupid; educated or uneducated.

Many remember Jackson less for his profound and thoughtful ideas on nervous function and disorder than in a more concrete context. His name is perpetuated whenever we speak of Jacksonian epilepsy, for the phenomenon of local onset of convulsions with orderly spread interested him for years, particularly when it proved to

result from 'coarse' but demarcated disease of one hemisphere. In his careful work on this topic Jackson characteristically gave credit to those who had preceded him, like Bravais and Todd. His studies led him to speculate on the nature of the more banal cases of 'idiopathic' epilepsy, and the attention of neurologists thus became diverted to the cerebrum from the medulla, the region of the brain hitherto under suspicion. Jackson is also remembered for his descriptions of what is now spoken of as temporal lobe epilepsy. He spoke of the 'intellectual aura' or the 'dreamy state', perhaps associated with hallucinations of taste and smell, and he proposed the term 'uncinate epilepsy'.

For many reasons Hughlings Jackson was not fully understood by his contemporaries, for his views were often unorthodox, well ahead of his time, and imperfectly communicated. Jackson was not a clear teacher or writer, and his desire to avoid overstatement, and to express his thoughts precisely, often defeated itself. He overloaded his text with footnotes and qualifying phrases, to the extent of obscurity. Few neurologists realised the supreme value of his message until after his death. Much of his modern appreciation has come about, ironically enough, through the intermediary of continental scientists like Pick, von Monakow, Freud and Sittig, who studied his writings though they were written in a foreign tongue, and then reinterpreted them back to us.

Jackson was anything but a stylist but he was masterly at the creation of arresting and felicitous phrases. Many of these have passed into the accepted grammar of neurology. It was he who spoke of superior and inferior levels of language, and identified swearing with 'propositions which were intellectually dead', being little more than 'detonating commas' or 'verbal missiles'. Milder interjections were dubbed 'feminine oaths'. To describe the verbal reiteration of some aphasiacs he spoke of 'recurring utterances' or 'barrel-organisms', which were really 'part of the rags and tatters of what had been the patients' speech' . . . 'like recurring decimals'.

Jackson's unique gifts to posterity comprised, therefore, adornments to the technique of neurology, to its philosophy, and to its vocabulary. Even today we may not have entirely grasped the magnitude of Jackson's ideas, for his intellectual life was a century ahead of his contemporaries. His doctrines remind us forcibly of Oscar Wilde's remark:

'. . . *There are works which wait, and which one does not understand for a long time; the reason is that they bring answers to questions which have not yet been raised; for the question often arrives a terribly long time after the answer.*'

THE 'FLOPPY' INFANT

JOHN N. WALTON, M.D., M.R.C.P.

Assistant Physician in Neurology, Royal Victoria Infirmary, Newcastle upon Tyne

The traditional concepts of hypotonic disorders in infancy need to be revised. These disorders are well grouped under: (1) infantile spinal muscular atrophy; (2) symptomatic hypotonia; and (3) benign congenital hypotonia. The clinical picture of the first (Werdnig-Hoffmann disease) as it actually occurs is described, the symptomatic hypotonias are reviewed, and the picture of benign congenital hypotonia made clear.

THERE can be few problems more perplexing to the paediatrician or neurologist than that of determining the nature of the pathological disturbance which is responsible for producing generalised hypotonia of the skeletal musculature in an infant. This clinical syndrome may be the result of a variety of diseases, some of which are grave, others benign, but all of which demand from the doctor the utmost in skill, tolerance and understanding in dealing with the affected child and his family. This group of disorders has been the graveyard of many a professional reputation and I am sure there can be few in the audience today who can claim never to have misdiagnosed a case of this nature. I have been interested in this problem for some time and was glad of your invitation to open your discussion this evening, but I must frankly admit that I have little that is new to contribute, and for me at least, infantile hypotonia poses many baffling questions. Accurate classification is still obscured by mists of confusion; some responsibility for this state of affairs must be laid at the door of those who have slavishly reproduced in current textbooks views which were first expressed concerning this group of disorders by eminent physicians shortly after the turn of the century.

Let us take, for instance, the syndrome of amyotonia congenita. Turn to a great many of the medical and paediatric textbooks in common use today and you will find that this condition is described as a generalised hypotonia of the skeletal muscles, present at birth, and showing a tendency to spontaneous improvement. On the other hand, infantile spinal muscular atrophy (Werdnig-Hoffmann disease) is commonly referred to as an acquired condition, beginning in the second six months of life, which gives progressive weakness and hypotonia of the muscles, develops in a previously strong and vigorous baby, and progresses rapidly to a fatal termination. This so-called 'classical' view certainly contains elements of truth, as we shall see in a moment, but rigid adherence to such a classification will lead to many diagnostic errors, with consequent distress both to those parents whose child deteriorates and dies after a reassuring interview, and to those whose infant recovers after months or years of waiting for the tragic end which had been forecast.

So many have been the cases in which a favourable prognosis has been belied by subsequent events that another school of thought has arisen, holding a view which implies that all cases of infantile hypotonia, beginning either at birth or subsequently, are due to an infantile disorder of anterior horn cells. This condition is most often progressive but may occasionally be arrested in infancy or may even fail to progress once the child is born and can then be regarded as a defect in development; some believe

* Read at the Royal Society of Medicine, Section of Paediatrics, on October 23, 1959.

that in such cases there may also be abnormalities of development of the upper motor neurone. If one follows this school of thought, then, in contrast to the 'classical' school, the diagnosis of amyotonia congenita carries a gloomy prognosis, implying the presence of a progressive spinal disease or at best an irreversible disorder of development. To apply this label and this kind of prognostic implication to all cases of infantile hypotonia is equally as unsound and just as productive of iatrogenic psychic trauma as would be slavish adherence to the 'classical' views.

To what can we attribute this confusion of thought and practice? First, as Greenfield (1957) recently pointed out, Oppenheim (1900), while doing a considerable service in drawing attention to those cases of infantile hypotonia which ran a benign course, subsequently discredited somewhat this benign syndrome of myatonia congenita, or as it is now called, amyotonia congenita, by diagnosing the condition in many 'floppy' babies whose disease later ran a progressively downhill course and was found to be a progressive infantile spinal muscular atrophy. Secondly, until recent years there has been relatively scant recognition of the fact that a great variety of muscular, skeletal, nervous and metabolic abnormalities can produce profound muscular hypotonia. Thirdly, there has been a serious lack of long-term follow-up studies of 'floppy' or hypotonic babies. Some of these deficiencies have recently been repaired by work in Scandinavia (Brandt 1950), in America (Ford 1952), in France (Arthuis 1954) and in this country, and much information of value has come to light. If we take as an example the series of 109 cases which I reviewed a few years ago (Walton 1956), this amply confirmed the view that the causes of infantile hypotonia, beginning at, or soon after, birth, can be grouped under three principal headings, which are:

1. Infantile spinal muscular atrophy.
2. Symptomatic hypotonia.
3. Benign congenital hypotonia.

Infantile Spinal Muscular Atrophy

It is now apparent that while this progressive degenerative disorder of the anterior horn cells frequently begins in the second half of the first year of life, as Werdnig and Hoffmann described, it is not uncommonly present at birth or begins shortly afterwards. Indeed, it remains the commonest cause of severe generalised hypotonia in infancy, no matter the age of onset, and it is probable that a similar process, beginning even earlier, in intra-uterine life, is responsible for a proportion of those babies which are born with widespread muscular atrophy and contractures, the condition which has been variously referred to as amyoplasia congenita or arthrogryphosis multiplex congenita. Some few cases of this type do, however, appear to be suffering from a congenital muscular dystrophy (Banker, *et al.* 1957), but in many more the condition is probably one of multiple fibrous contractures without evidence of disease in the muscle fibres or motor neurones and it is possible that excessive intra-uterine pressure may play a part in its causation. The condition which Löwenthal (1954) has referred to as familial myosclerosis is clearly closely-related.

The tragic clinical picture of infantile spinal muscular atrophy is one which must be only too painfully familiar to all paediatricians and neurologists. While the course and presentation may be variable, depending upon the age of onset and rapidity of progress of the disease, the appearance of the established case is characteristic and unforgettable. The baby lies virtually immobile with arms abducted at the shoulder and lying on either side of the head and with the legs abducted and externally rotated at the hips. Only minimal movement of the more peripheral muscles is possible, those lying more proximally being almost completely paralysed, and respiration is almost exclusively diaphragmatic, with recession of the lower thoracic cage occurring with each shallow breath. On being lifted the baby droops limply and will seem to slip through the hands, while bizarre postures and excessive passive

movements of the paralysed limbs are possible. All these features may be seen in a baby which was at first sight healthy and well-nourished until at some stage the parents observed that the child's movements were unusually weak or restricted or that its limbs were limp and 'floppy'. Almost the only prospect which can now be predicted is one of slow insidious deterioration with subsequent bulbar paralysis. Repeated respiratory infections occur at progressively shorter intervals, until finally, with a cough which is increasingly feeble, the infant's resistance, bolstered by all the known antibiotics, is inadequate to counter an infection more insistent than the last. True, there are a small proportion of cases in which the disease process appears to become arrested, but those who have seen pitiably deformed and helpless children surviving from this disease even into adolescence must wonder whether those dying in infancy were not more fortunate.

More tragic still is the fact that this condition, being genetically-determined and probably due to an autosomal recessive gene, will afflict at least one in four children in any sibship and often considerable more, so that one may witness, as I have done, three consecutive children of a young and vigorous couple succumbing to this affliction. Can it be wondered that most intelligent parents, being told, as they must, of the risk involved, refuse to contemplate further pregnancies?

Symptomatic Hypotonia

The number of pathological disorders in infancy which can produce some degree of hypotonia of the skeletal muscles is legion and I do not propose to catalogue them here. The cases of this type can be said to fall into five principal groups.

First we have the muscular and neuromuscular disorders, such as muscular dystrophy, polymyositis, myasthenia gravis and infantile polyneuritis. Hypotonia and excessive mobility of the joints is uncommon in progressive muscular dystrophy but it can, rarely, be the presenting feature in the few cases which begin in the first year of life; a positive family history may help in diagnosis, and subsequently the characteristic waddling gait and muscular pseudohypertrophy become evident. Muscle biopsy may establish the diagnosis, as it usually will in those rare cases of polymyositis which develop in infancy and in which signs of constitutional disease are absent or unobtrusive. Myasthenia gravis must always be borne in mind, particularly if there is ptosis and other evidence of weakness of the cranial musculature, when an injection of edrophonium or prostigmine may clinch the diagnosis. Polyneuritis is comparatively rare in infancy but in many such cases the limb weakness develops comparatively rapidly in a previously healthy child and the cerebro-spinal fluid protein is found to be raised.

Secondly we have a group of cerebral disorders, including, for example, the brain-injured child, whose limbs may be found to be weak and hypotonic following on a traumatic labour or a prolonged period of neonatal apnoea. Hypotonia and an undue mobility of the limbs is also relatively common in those cases of cerebral palsy which have a flaccid diplegia and in which the development of motor skills is considerably delayed; in these cases athetotic posturing of the limbs or ataxia of cerebellar type may be observed, even at a relatively early age. One of the most difficult of all diagnoses to make in this group is that of mental defect, which may call for all the doctor's skill and experience in assessing the infant's behaviour against a background of known variations in normal development. Many of us can remember placid and apparently contented babies whose generally limp and hypotonic condition led to serious consideration of a diagnosis of Werdnig-Hoffmann disease, until it became clear that their delay in development of motor activity was accompanied by a parallel defect in intellectual accomplishment. Comparative rarities such as cerebral lipidosis and kernicterus, which may also produce a hypotonic child, must also be mentioned, but fits, and myoclonus

particularly, are usual in lipidosis, while severe deafness, and rigidity rather than hypotonia of the limbs are often seen in kernicterus.

A great many of the disorders giving rise to symptomatic hypotonia fall into the third group—that of nutritional and metabolic disorders. Usually in these cases the concomitant fever and wasting or other evidence of systemic disease will reveal the nature of the primary disorder of which impaired muscular activity and reduced tone are but a symptom. It is, however, remarkable how long these features may persist after a febrile illness, while it can be relatively easy to overlook the bulky stools and flattened buttocks of the infant with coeliac disease or the irritability and other revealing signs in disorders such as hypercalcaemia, scurvy and even chronic urinary infection with nephrocalcinosis. Rickets, once a common cause of this clinical picture, is no longer to be seriously considered in Great Britain, save in children with renal disease, while the early diagnosis of cretinism is a diagnostic hurdle so well recognised that it seems to be accepted in paediatric circles as a common exercise for the fledgling specialist and as a justifiable means of demonstrating the paediatrician's superiority over lesser mortals. Another relatively rare but important cause of profound generalised muscular hypotonia is glycogen storage disease. Sometimes the liver and spleen are enlarged but more often the disorder of glycogen storage is confined to the muscles themselves and diagnosis must then depend upon histochemical examination of a specimen of muscle removed by biopsy.

In yet a fourth group of cases we have a variety of skeletal disorders, including the child with long, spidery fingers, high arched palate, slender muscles and mobile limbs—the subject of arachnodactyly. The infant with osteogenesis imperfecta, too, who soon demonstrates his liability to fractures on minimal trauma, may have relatively immobile limp and 'floppy' limbs. By contrast, in the child with congenital laxity of the ligaments, in whom there is also as a rule a clear family history (many relatives may have been contortionists), spontaneous movement of the limbs is vigorous and full but yet a remarkable degree of passive mobility at the joints is possible.

Finally we come to a fifth and miscellaneous group of conditions, including disorders as diverse as congenital heart disease and other congenital abnormalities, many of which may be associated with moderate muscular hypotonia, and spinal cord birth injury, in which the flaccid paralysis of the lower limbs may be erroneously attributed to a disorder of the lower motor neurone. The pseudoparalysis of congenital syphilitics is rarely seen nowadays but the relative immobility resulting from bilateral congenital hip dislocation is sometimes mistaken for muscular hypotonia.

This list is a formidable one, and even so there are many omissions. I hope I have brought out the importance of bearing in mind the possibility that the disease responsible for a particular infant's hypotonic state can sometimes be far removed from the lower motor neurone and that assiduous clinical enquiry may bring its nature to light.

Benign Congenital Hypotonia

This is probably the most interesting and yet the most perplexing group of all. These children show impaired spontaneous mobility of the limbs, delay in physical development, and a generalised hypotonia of the musculature, which can be remarkably profound and is apparent soon after birth. In severe cases there is even weakness of the respiratory muscles. After a variable period of apparent standstill, steady improvement is the rule. In a proportion of cases of this type recovery is complete, though the child may have slight residual weakness or hypotonia even into early adolescence. Another group of cases improve up to a point but always retain some degree of disability and show a curiously slender skeletal musculature, often combined with some residual hypotonia, which persists throughout life.

Evidence of skeletal abnormalities and metabolic disturbances are lacking and although electromyography will give a suggestively myopathic pattern in some cases, muscle biopsy specimens, when studied with conventional techniques, are almost invariably normal. It is possible that the newer histochemical techniques will yield important dividends if applied to muscle removed from cases of this type, and studies of terminal innervation are also likely to be profitable, but it is too early to say whether the interesting results of pioneer studies of this type (Shy and Magee 1957, Coërs and Pelc 1954, Woolf 1957) are likely to be generally applicable to all cases of this nature.

These cases are much less common than those of spinal atrophy and symptomatic hypotonia, but I have now seen a total of 28 patients of this type (Walton, 1957*a* and *b*). All of them were limp, 'floppy' babies, and virtually all of them walked later than the normal age—some not until they were five years old. In each case there was weakness and hypotonia, particularly of proximal limb muscles; half the patients recovered completely, while half showed some improvement but never became normally powerful, and some weakness of proximal muscles, combined with a general smallness of the musculature, persists. In general those cases showing incomplete recovery were more profoundly weak and hypotonic at the beginning than those who recovered completely.

I have little doubt that this condition which I have called 'benign congenital hypotonia' is essentially the same as myatonia congenita or amyotonia congenita as described by Oppenheim (1900). But in view of the widespread confusion which exists concerning the significance of 'amyotonia congenita' as a diagnostic label I feel that it is better discarded and the new name substituted. Furthermore, there seem to be good reasons for supposing that Turner's (1940, 1949) 'benign congenital myopathy' and Krabbe's (1946) 'congenital universal muscular hypoplasia' are very closely related to, if not identical with, those cases of benign hypotonia which recover incompletely.

Leaving aside all arguments concerning terminology, it is, however, apparent that this is an important group of cases, in view of the relatively favourable prognosis. Of its aetiology we understand very little. Is it a disorder of prenatal neuromuscular development which can sometimes be compensated for by postnatal maturation processes? Is it a biochemical disorder of the muscle fibre? Does it originate in a disturbance of function in the pathways concerned with muscle tone, either peripherally in the muscle spindles or centrally in the spinal cord? Or are we in fact dealing with a variety of different pathological entities which can produce this clinical syndrome? We cannot even begin to answer these questions today and I fear it will be many years before they are elucidated. For the moment we must content ourselves as clinicians with recognising the existence of this benign syndrome and with devising clinical and other means of distinguishing it from the other causes of infantile hypotonia.

One cannot but suspect that, particularly in the cases which recover incompletely and have weak and slender muscles throughout life, we may be dealing with a variety of diseases which nevertheless show a remarkable uniformity of clinical presentation. For instance, I am sure that arachnodactyly can produce this clinical picture. Furthermore, Shy and Magee (1957) have recently described a family of cases of this type in which biopsy revealed muscle fibres which were larger than normal and which contained, in transverse section, a central core of closely-packed myofibrils which stained blue, instead of the normal purple, with Gomori's trichrome stain. I have looked for this change in several of my cases but have not yet found it, and up to the present this family with so-called 'central core disease' appears to be unique.

In another recent paper, Greenfield, Cornman and Shy (1958) have studied the value of muscle biopsy in differential diagnosis in 'floppy' infants. They accept the

existence of a syndrome of benign infantile hypotonia which recovers completely and in which muscle biopsy specimens are normal, but feel that many of the patients which show incomplete recovery are in fact suffering from a congenital progressive muscular dystrophy. This view has not been confirmed by my observations, and it is the complete absence of clinical progression and of recognisable histological change in the muscles, even in the cases whose muscles remain small and weak, which has led me to disagree with Turner's (1949) suggestion that this condition is a congenital myopathy, if by myopathy one is to imply that it is a primary muscular disease akin to muscular dystrophy.

Krabbe (1958) may be more nearly correct in describing the condition as one of universal muscular hypoplasia, but it is clear that this problem will remain unanswered so long as we depend upon histological techniques which can distinguish so limited a range of pathological changes in muscle.

Differential Diagnosis

There can be few clinical syndromes in which accurate diagnosis is more important for prognostic reasons than in the hypotonic child. If a diagnosis of spinal muscular atrophy is established, then the outlook is grave; if symptomatic hypotonia, the prognosis is that of the underlying disease, which may require specific treatment; while if benign hypotonia is diagnosed, improvement can be confidently predicted, though it may not be apparent for some time whether the child will recover completely or whether some disability will persist.

We have already considered some of the points which may help in the differential diagnosis of symptomatic hypotonia; in this condition the reduction in muscular tone and the excessive degrees of passive movement possible are rarely so great as in cases of spinal atrophy or even of benign hypotonia. The virtually immobile child with spinal muscular atrophy, limp and unresisting, with shallow rapid respirations and indrawing of the lower ribs, presents a clinical picture which is virtually diagnostic, and observation of fasciculation in the tongue will clinch the diagnosis. In benign congenital hypotonia, by contrast, though the child may be remarkably limp and 'floppy', there is always some spontaneous movement of the limbs, the tendon reflexes are less profoundly depressed and respiratory difficulty is very rare. Despite these clinical pointers, a final decision in any individual case can be very difficult to reach. Sometimes the passage of a few weeks or months and the subsequent improvement or deterioration in the patient's condition will make the position entirely clear, but in other instances, particularly when the demands of the naturally anxious parents for a definite answer are pressing, it may be justifiable to carry out a number of ancillary investigations in hospital.

Of these, soft-tissue X-ray films can help in demonstrating the profound muscular atrophy of the spinal disease, while haematological and biochemical tests may be required to exclude certain causes of symptomatic hypotonia. More important, however, are electromyography and muscle biopsy. If the electromyogram gives a myopathic pattern, spinal atrophy can be excluded with reasonable confidence; muscle biopsy is even more useful, for diagnostic changes are found in practically all cases of the spinal disease and a normal result is greatly in favour of benign or symptomatic hypotonia. Perhaps newer histochemical techniques and the use of the electron microscope will tell us even more.

Despite the value of these investigations in many cases, there are others in which the results are equivocal and we must resort to repeated clinical observation over a period of months with all the distress which this course necessarily entails for both the patient's family and the doctor. Indeed, diagnosis in the hypotonic child is never an easy matter, but the very existence of the benign cases gives some hope that all may not necessarily be quite so gloomy as it first appeared. I realise that in connection

with benign hypotonia I have raised more questions than I have answered but we may hope that further studies of these interesting cases will throw new light on the whole problem of infantile hypotonia, a problem which is certainly one of the most difficult but at the same time one of the most challenging to clinical ability in the field of medicine.

SUMMARY

The clinical syndrome of generalised hypotonia of the skeletal musculature occurring in infancy may be due to a variety of diseases, some of which are grave, others benign, but all of which present difficult diagnostic problems.

The first broad group of cases which show the clinical picture are those of infantile spinal muscular atrophy. In this condition, hypotonia and muscular weakness may be present at birth or may begin postnatally in the first two years of life. The disease is steadily progressive, producing widespread paralysis, and most affected infants die from respiratory infection within a few months of the onset, though some survive, pitiably deformed, for several years.

In a second large group of cases the hypotonia of the skeletal muscles is but one feature of an illness which may or may not be producing pathological changes in the central or peripheral nervous system. This large group of diseases which produce symptomatic hypotonia includes muscular and neuromuscular diseases (muscular dystrophy, polymyositis, myasthenia gravis and infantile polyneuritis), cerebral disorders (flaccid diplegia, mental defect, lipidosis, kernicterus), nutritional and metabolic disorders (hypercalcaemia, scurvy, chronic urinary infection, malabsorption syndrome, cretinism, glycogen storage disease), skeletal disorders (arachnodactyly, congenital laxity of the ligaments, osteogenesis imperfecta) and miscellaneous conditions (congenital heart disease, spinal cord birth injury, etc.).

The third main group is that of benign congenital (or infantile) hypotonia, in which hypotonia may be profound and muscular weakness considerable, and in which there is a pronounced delay in reaching physical milestones (sitting up, walking, etc.). Some such patients recover completely in time, but others show a degree of muscular hypotonia, and small, slender skeletal muscles, throughout life. This condition corresponds in many respects to Oppenheim's 'amyotonia congenita', Turner's 'benign congenital myopathy' and Krabbe's 'congenital universal muscular hypoplasia'. However, the term amyotonia congenita should be discarded except as a descriptive title including all causes of infantile hypotonia, for many doctors use this term to describe cases of infantile spinal muscular atrophy of early onset.

The differential diagnosis between these many causes of hypotonia in infancy is discussed and the value of electromyography and muscle biopsy is considered.

Despite all ancillary aids, accurate diagnosis often depends on repeated observation of the child over several months.

RÉSUMÉ

Les Nourrissons 'Mous'

Le syndrome clinique de l'hypotonie généralisée de la musculature du squelette, dans la première enfance, peut être dû à une variété de maladies. Quelques unes sont graves, d'autres bénignes mais toutes présentent des problèmes de diagnostic ardus.

Le premier et large groupe de cas ayant le tableau clinique est celui de l'atrophie musculaire spinale infantile. Ici, l'hypotonie et la faiblesse musculaire peuvent soit exister à la naissance, soit se manifester après, au cours des deux premières années de la vie. La maladie augmente régulièrement. Elle déclanche des paralysies étendues. La plupart des nourrissons meurent d'infection respiratoire, quelques mois après le début. Quelques rescapés, pitoyablement déformés, survivent plusieurs années.

Dans un vaste second groupe, l'hypo-

tonie des muscles du squelette n'est qu'un des signes d'une maladie capable ou non de produire des modifications pathologiques du système nerveux central ou périphérique. Ce vaste groupe de maladies génératrices de l'hypotonie symptomatique comprend: des maladies musculaires et neuromusculaires (myopathie, polymyosite, myasthenia gravis et polynévrite infantile); des troubles cérébraux (diplégie flasque, arriération mentale, lipidose, ictère nucléaire); des troubles de la nutrition et du métabolisme (hypercalcémie, scorbut, infection urinaire chronique, syndrome de malabsorption, crétinisme, polycorie glycogénique); des troubles du squelette (arachnodactylie, laxité congénitale des ligaments, osteogenesis imperfecta) et des états divers (cardiopathie congénitale, lésion médullaire obstétricale, etc.).

Le troisième groupe principal de cas est celui de l'hypotonie bénigne congénitale (ou infantile). L'hypotonie peut y être profonde et la faiblesse musculaire considérable. Il y a un retard prononcé dans les différentes étapes du comportement physique (station assise, marche, etc.). Quelques malades guérissent complètement avec le temps, cependant que d'autres gardent toute leur vie un certain degré d'hypotonie musculaire et une musculature menue et fusiforme. Cet état correspond par bien des aspects à la myotonie congénitale d'Oppenheim, à la 'myopathie congénitale bénigne' de Turner et à 'l'hypoplasie musculaire congénitale généralisée' de Krabbe.

Cependant il faut se garder d'employer le terme de myotonie congénitale autrement que pour situer sous ce titre signalétique toutes les causes d'hypotonie infantile; en effet de nombreux médecins usent de cette expression pour décrire les cas d'atrophie musculaire infantile de la moelle à début précoce.

Le diagnostic différentiel entre ces nombreuses causes d'hypotonie infantile est discuté; la valeur de l'électromyographie et de la biopsie du muscle est étudiée. En dépit de tout examen complémentaire, un diagnostic exact dépend souvent d'observations répétées de l'enfant s'échelonnant sur plusiers mois, jusqu'à ce que le diagnostic se clarifie.

ZUSAMMENFASSUNG

Schlaffe Sauglinge

Das klinische Bild der in der ersten Kindheit auftretenden generellen Hypotonie der Sklelettmuskulatur kann durch verschiedene Krankheiten hervorgebracht werden. Einige von ihnen sind gefährlich, andere harmlos, aber alle stellen schwierige diagnostische Probleme.

Die erste weite Gruppe von Fällen, die dieses klinische Bild aufweisen, ist die der infantilen Spinalen Muskelatrophie. Hier können Hypotonie und Muskelschwäche entweder seit der Geburt vorhanden sein, oder nachher, während der zwei ersten Lebensjahre anfangen. Die Krankheit nimmt regelmässig zu, indem sie weit ausgebreitete Lähmungen hervorbingt. Die meisten Säuglinge gehen einige Monate nach dem Ausbruch an respiratorischen Infektionen zugrunde. Einige, jämmerlich verunstaltet, bleiben noch mehrere Jahre am Leben.

In einer zweiten weiten Gruppe von Fällen ist die Hypotonie der Skelettmuskulatur nur eines der Zeichen einer Krankheit, die ab und zu pathologische Veränderungen des zentralen oder peripheren Nervensystems erzeugen kann. Diese ausgedehnte Gruppe von Krankheiten, die Symptomatische Hypotonie erzeugen können, setzt sich aus muskulären und Neuromuskulären Erkrankungen (Muskeldystrophie, Polymyositis, Myasthenia gravis und infantile Polyneuritis), zerebralen Störungen (schlaffe Diplegie, Intelligenzdefekt, Lipidosis, Kernicterus), Ernährungs—und Stoffwechselstörungen: (Hypercalcaemia, Skorbut, chronische Infektionen der Harnwege, malabsorbtion Syndrom, Kretinismus, Glykogen-Aufspeicherungsstörung) Skelettkrankheiten (Arachnodaktylie, Kongenitale Schlaffheit der Ligamente, Osteogenesis imperfecta) und verschiedenen anderen Zuständen (Angeborene Herzfehler, Geburtschädigungen des Rückenmarks, usw.) zusammen.

Die dritte Hauptgruppe ist die der harmlosen kongenitalen (oder infantilen) Hypotonie, die durch tiefe Hypotonie und erhebliche Muskelschwäche gekennzeichnet sein kann, und in der es eine stark ausgeprägte Verspätung der körperlichen Entwicklung gibt: (Sitzen, Gehen, usw.) Einige Kranken genesen vollständig mit der Zeit, während andere ihr ganzes Leben lang eine gewisse Muskelhypotonie und eine kleine und spindelförmige Skelettmuskulatur behalten. Dieser Zustand entspricht in mancher Hinsicht der Myotonia congenita von Oppenheim, der harmlosen kongenitalen Myopathie von Turner und der kongenitalen generellen Muskelhypoplasie.

Doch darf der Name Myotonia congenita nicht anders angewendet werden, als wie eine Bezeichnung, die alle Ursachen der infantilen Hypotonie umfasst. Viele Arzte gebrauchen nähmlich disen Ausdruck, um Fälle frühinfantiler spinaler progressiver Muskelatrophie zu beschreiben.

Die Differentialdiagnose zwischen diesen zahlreichen Ursachen der infantilen Hypotonie wird erörtert und der Wert der Elektromyographie und der Muskelbiopsie wird erwägt. Trotz aller Hilfsmethoden, hängt eine genaue Diagnose oft von wiederholten Beobachtungen des Kindes ab, die sich auf mehrere Monate ausstrecken, bis die Diagnose klar wird.

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MUSCLE BIOPSY IN THE DIAGNOSIS OF THE "FLOPPY BABY": INFANTILE HYPOTONIA*

A. L. WOOLF, M.D., M.R.C.P.
Midland Centre for Neurosurgery, Smethwick, Staffs

New techniques of muscle biopsy, by demonstrating the intramuscular nerve-endings, can help in the diagnosis of 'floppy babies', by providing evidence of disease of the spinal cord or peripheral nerves not demonstrable with classical methods.

THE object of this paper is to set out in the light of personal experience the value of muscle biopsy in the diagnosis of the 'floppy' infant. I shall deal first with the orthodox muscle biopsy, in which classical staining methods only are used, and shall then consider the value of the special muscle biopsy, using methods which demonstrate the motor and sensory nerve endings, including the motor end-plates and the synapse itself. My personal experience comprises 25 cases where the patient was a 'floppy' infant or child, and these cases were taken from a series of just over 300 muscle biopsies in which the special methods were employed.

The greatest value of the orthodox muscle biopsy lies in its ability to demonstrate beyond doubt the presence of a lesion of the lower motor neurone. The characteristic grouping of small and normal sized muscle fibres is too well known to need further description. It is not possible by studying these fibres in the ordinary way to deduce the cause of the lesion or whether it is progressive or not. Thus the appearances are the same after poliomyelitis or infantile polyneuritis, or in the course of Werdnig-Hoffmann disease.

Walton (1956) has listed, under the heading of symptomatic hypotonia, a number of other conditions in which muscle biopsy might be expected to reveal structural changes in the muscle, including progressive muscular dystrophy, polymyositis, and glycogen storage disease. I have never seen a case of any of these diseases presenting as a 'floppy' infant, but Walton has seen two children who had been hypotonic in infancy and who, late in the first or early in the second decade, showed the characteristic features of the Duchenne type of muscular dystrophy. Walton has also seen at the age of 8 years a single case of polymyositis in which the patient had shown weak, limp muscles soon after birth. In this case muscle biopsy, presumably performed at the age of 8, revealed a chronic myositis. Bovet (1936) had a single similar case. However, these primarily muscular disorders must be regarded as rare causes of floppiness in infancy, though, of course, it is right to bear them in mind when studying muscle biopsy specimens.

* Opening paper at a meeting of the Section of Paediatrics of the Royal Society of Medicine, October 23, 1959. This study was aided by a grant from Muscular Dystrophy Associations of America, Inc.

Thus we may expect the most frequent positive biopsy finding in 'floppy' infants to be neurogenic atrophy, and this will most commonly be a manifestation of Werdnig-Hoffmann disease.

Special Techniques

We may now consider the special biopsy techniques which demonstrate the intramuscular nerve endings. The details of the methods employed to localise the innervation zone, the techniques of vital staining with methylene blue, and the histochemical demonstration of cholinesterase are set out in the book I have written with Dr. Coërs (Coërs and Woolf 1959), and I will not refer to them here. By their means we can detect pathological changes in the axons and end-plates, even in otherwise normal-looking muscle. These changes are of considerable diagnostic and prognostic value in differentiating between the different forms of neurogenic muscular atrophy which are indistinguishable in biopsy specimens studied by classical methods.

Thus, we have observed by means of these methods in six cases a characteristic change in the nerve-endings which we interpret as a 'dying back' of the neurone (Coërs and Woolf 1959). This appearance seems to be diagnostic of an abiotrophy of the lower motor neurone. The first five of these cases presented clinical features of Werdnig-Hoffmann disease, but the patients are either still alive or have not been studied by thorough post-mortem neuropathological methods.

The last case is of particular interest since it shows the importance of establishing the diagnosis in this way. It concerned a 'floppy' infant showing the biopsy picture of 'dying back' of the neurone but in which there was not only anterior horn cell degeneration but also degeneration of the posterior and lateral columns of the spinal cord and almost total atrophy of the cerebral white matter with severe hydrocephalus. A brother had died of a similar illness and also showed marked ventricular dilation. This case makes it clear that not every case of abiotrophy of the anterior horn cells commencing in infancy is to be regarded as an example of Werdnig-Hoffmann disease, and therefore that this disease cannot be diagnosed with complete certainty during life.

Where Classical Staining Shows No Abnormality

We must now consider the cases in which by using classical staining methods the muscle appears normal. Essentially these seem to fall into three groups: (1) cases where even with full post-mortem investigation there is no demonstrable abnormality in the nervous system; (2) cases where there is a demonstrable abnormality, but it does not affect the lower motor neurone; and (3) cases where the lower motor neurone is affected but the lesion cannot be demonstrated by classical staining methods.

Group 1 will include cases resembling Oppenheim's amyotonia congenita and those examples of Walton's benign congenital hypotonia in whom the intramuscular nerve endings are shown to be normal by the new techniques, and also mentally defective children. Cases can be legitimately placed in this group only if thorough post-mortem neuropathological examination has shown that the nervous system is intact, and it seems certain that if such examinations are performed and new biopsy techniques are applied, this group will progressively diminish. Thus in regard to infants with mental retardation, one may envisage—and I have studied one such case (Fig. 1)—in cerebral birth injury or other gross disease of the brain that the anterior horn cells may have come within the ambit of the destructive process. This would provide a much-wanted explanation for the commonplace but puzzling association of mental defect and muscular hypotonia in those cases often referred to as atonic Little's diplegia. It would at the same time, of course, place them in Group 3, which would thus be enlarged at the expense of the dwindling Group 1. However, although in one 'floppy' infant who later was shown to have epilepsy and mental defect,

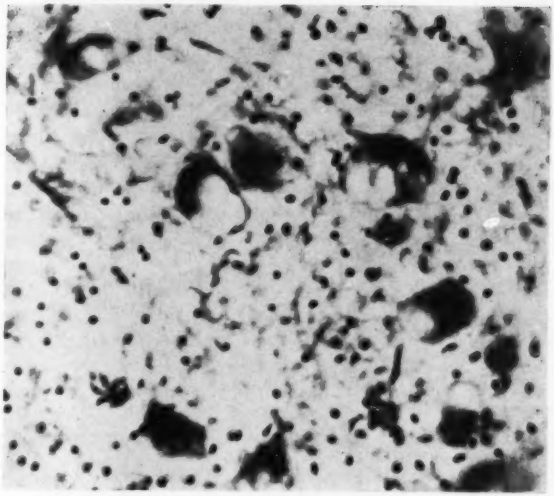


Fig. 1. 6th cervical segment of spinal cord from infant aged 5 weeks with bilateral cerebral softenings from anoxia during birth. Anterior horn cells show vacuolar degeneration with accompanying neuroglial proliferation. (Nissl x 510)

the end-plates were poorly formed, in most of the cases of mental deficiency with hypotonia from which I have examined muscle biopsies, including one mongol, the motor and sensory nerve endings were normal. It seems, therefore, that in most cases the hypotonia of mental deficiency has its basis in disordered function of the reflex pathways or in some more subtle disorder of the end-plate than we have yet been able to demonstrate.

In the second group of cases, although the biopsy shows normal muscle, there is an abnormality in the nervous system not affecting the anterior horn cells. This group is important since it would be most unfortunate to give a good prognosis on the basis of a normal muscle biopsy when the child's immobility is due to progressive disease of the brain or spinal cord. Of the many diseases which might produce this situation we may cite one personal case in

Fig. 2. Biopsy specimen from peroneus brevis of child aged 7 whose muscles had been hypotonic since birth. The fasciculi which are composed of normal-sized muscle fibres are widely separated by fat. There were no atrophic muscle fibres (same case as Figs. 204 and 205 in Coërs and Woolf 1959) (H & E x 80)

which a normal muscle biopsy was obtained from an infant who proved later to have a leukodystrophy of the familial type.

It is in the third group that one feels most strongly the value of the new methods in demonstrating abnormalities in the nerve endings. In these cases the muscle with haematoxylin and eosin staining appears normal or there is at the most an increased amount of fat between the fasciculi, so that

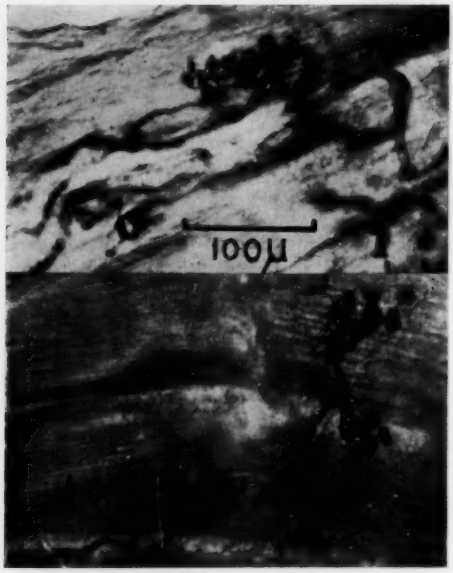
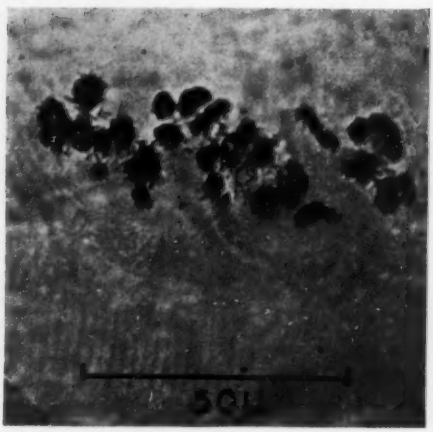


Fig. 3. Boy aged 7 years, originally presented as amyotonia congenita. Some end-plates are very large and elaborate: others are very small and simple. Probably sequel to infantile polyneuritis. Lower photograph shows innervation of two muscle fibres by one axon.
(Vital staining with methylene blue)

if one did not know that muscle fibres could completely disappear after denervation one might doubt whether a lesion of the lower motor neurone had in fact occurred (Fig. 2). Any such doubts should, however, be dispelled by the strikingly abnormal end-plates and terminal axons (Figs. 3–6). Vital staining may show that the axons are reduced in number, irregularly thickened, or exhibit excessive and disorderly branching. In other cases there is collateral reinnervation and great irregularity in the size and complexity of the end-plates. These cases are not usually examples of Werdnig-Hoffmann disease as classically described. Typically they have been hypotonic since birth, are not deteriorating, or may even be improving. Some of them may have had a polyneuritis during infancy or even in intrauterine life; others may be *formes frustes*, or extremely slowly progressive cases of Werdnig-Hoffmann disease. Yet others (Fig. 1) may have developed anterior horn cell lesions as a result of birth anoxia, but this is speculation. What is needed is thorough post-mortem studies of such cases. This will, of course, only rarely be possible, because of the benign prognosis. It is, therefore, all the more important to take full advantage of what opportunities present. I have no doubt that some of Walton's cases of benign congenital hypotonia would fall into this group if their intramuscular nerve endings were brought under scrutiny. In one of my cases the changes were very similar to those described by Coërs as immature. I contend that these changes may well represent poor regeneration after an arrested denervation process,

Fig. 4. Same case as Fig. 3. Cholinesterase preparations show subneural apparatus corresponding to very elaborate end-plate in Fig. 3 (upper).
(Koelle's technique modified by Couteaux)



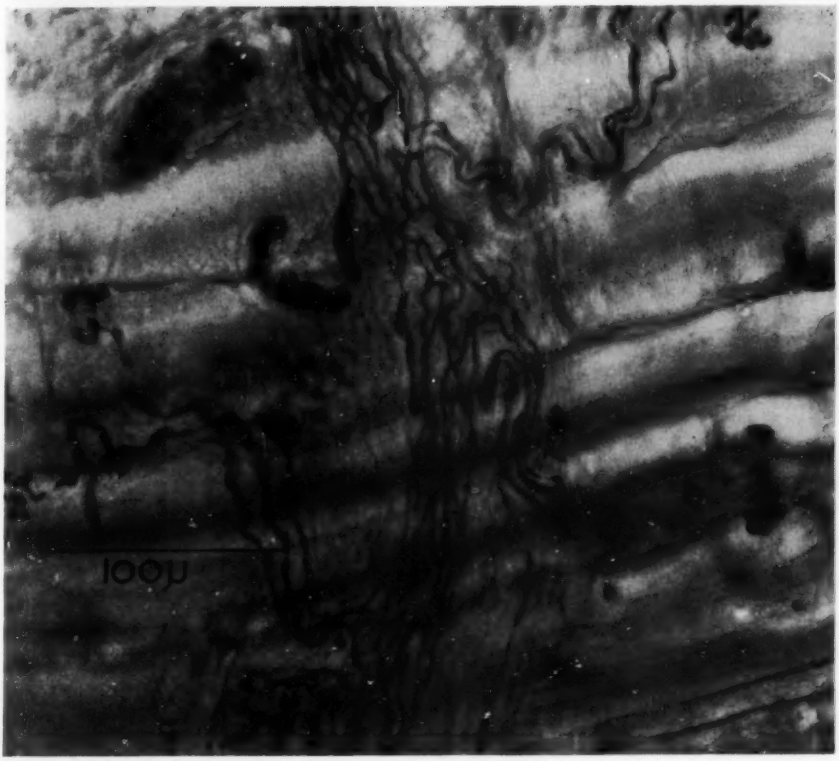


Fig. 5. Normal intramuscular nerve bundle with end-plates, showing comparatively uniform structure of the latter, from boy aged 7 years.
(Vital staining with methylene blue)

and I have yet to be convinced that cases in which muscle tone appears to develop late are expressions of delayed but normal development of the nerve endings.

Value of Electron Microscopy

Electron microscopy has several advantages over light microscopy in addition to its greatly increased powers of magnification and resolution. By means of electron microscopy, myelin sheaths, axons, mitochondria, cytoplasm and nuclei of cells, collagen fibres and other structures can all be seen in a single section instead of a different preparation being required for each element to be studied (Fig. 7). In addition, electron microscopic technique is less capricious than vital staining. This especially applies to degenerating end-plates, and in one of the muscle biopsies from cases of Werdnig-Hoffmann disease which we have studied by this means end-plates were clearly demonstrated, while in vitally stained preparations the fine beaded sprouts appeared to end without forming an end-plate. Electron microscopic preparations also reveal fine beaded unmyelinated sprouts which are invisible with the light microscope (Fig. 7).

Finally, we may consider whether there may not be changes at the synapse itself which are too subtle to be seen with the

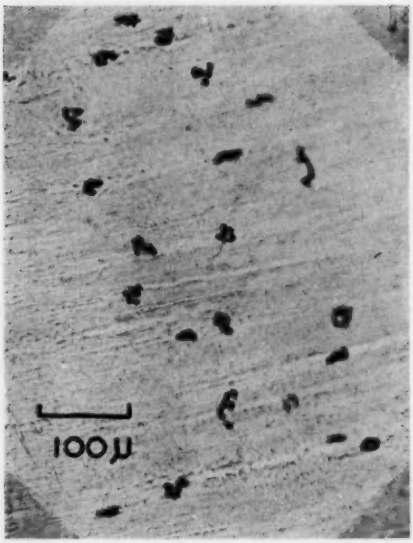


Fig. 6. Normal subneural apparatuses of end-plates in boy aged 7 years.
(Koelle's technique modified by Couteaux)

light microscope even after vital staining. Such changes might, for example, underlie the disorder of neuromuscular transmission found in myasthenia gravis.

Here again our recent studies with the electron microscope are of some interest, and I will conclude this paper by illustrating the appearance of the end-plates as studied by this instrument. You will observe that the terminal expansions are no longer to be considered as uniform structures, but as containing mitochondria and synaptic vesicles probably composed of acetylcholine (Fig. 8); about a million molecules is the suggested content of each vesicle (Acheson 1948). We may further see that there is a compound synaptic membrane which we believe we have demonstrated in man for the first time (Bickerstaff *et al.* 1960). Disorder of this membrane might well explain some of the temporary or early disorders and delays in development of tone. But this is for the future; at present we must content ourselves with the prospect of important new discoveries as this wonderful new instrument is put to increasing good use.

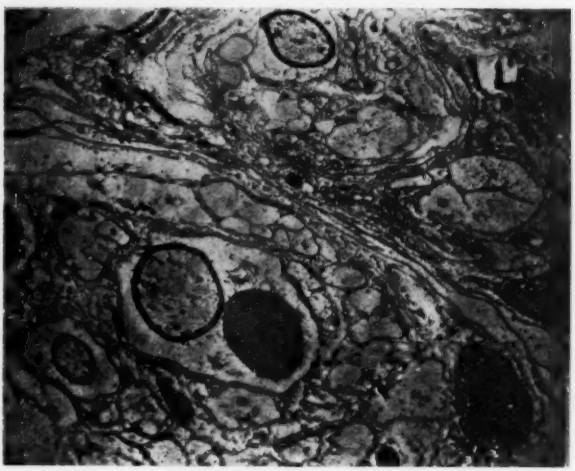


Fig. 7. Electron microscope preparation of intra-muscular nerve bundle in case of Werdnig-Hoffmann disease. There are 3 myelinated nerve fibres, with unusually slender myelin sheaths enclosing axons showing several mitochondria The largest myelinated fibre lies within the cytoplasm of a Schwann cell, beside the latter's nucleus. There are numerous unmyelinated axonic sprouts partially surrounding the Schwann cell. (Osmic acid fixation x 4,000)

ACKNOWLEDGEMENTS

Figs. 3–6 have previously been published in 'The Innervation of Muscle', by C. Coërs and A. L. Woolf and are repro-

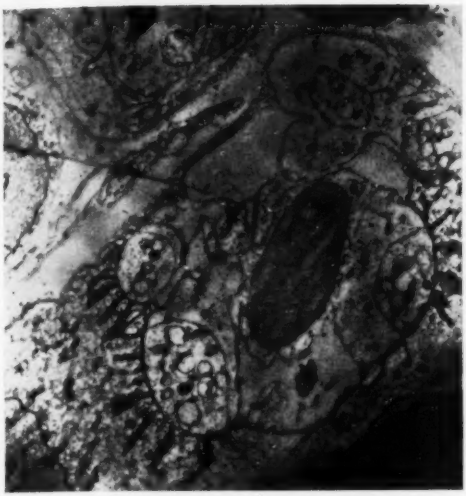


Fig. 8. Electron microscope preparation of motor end-plate from a case of peripheral neuropathy following Wernicke's encephalopathy. Three terminal expansions of the neural component of the end-plate are shown. They contain large vesicular mitochondria and fine grey synaptic vesicles probably containing acetylcholine. They are formed on the muscle side by the synaptic membrane which dips into the petal-like junctional folds.
(Osmic acid fixation x 5,700)

duced with the kind permission of Messrs. Blackwell Scientific Publications Ltd.

The case illustrated in Fig. 1 was studied in Dr. M. Bodian's department at the Hospital for Sick Children, Great Ormond Street, London.

I am grateful to Dr. D. R. Humphreys for permission to refer to the case mentioned in the legend to Fig. 8.

SUMMARY

The intramuscular nerve endings in biopsy specimens from 25 'floppy' infants were studied by vital staining, histochemical demonstration of cholinesterase and electron microscopy. The cases can be divided into two main groups:

(*a*) Those in which the biopsy specimen studied by classical staining methods appears abnormal. The most frequently encountered abnormality is the well-known atrophy of muscle fibres by groups, indicating degeneration of the lower motor neurone; in 'floppy' infants the degeneration is most commonly due to Werdnig-Hoffman disease. The appearance of the intramuscular nerve endings in this disease as shown by the new methods is very characteristic and of considerable diagnostic value.

(*b*) Cases where with classical staining methods the muscle biopsy appears normal. These in turn may be divided into three subgroups:

(1) Cases where even with air encephalograms and full neuropathological post-mortem examinations there is no demonstrable abnormality in the nervous system—these cases appear to correspond to Oppenheim's amyotonia congenita and some of Walton's benign congenital hypotonia cases. It is certain that as the proportion of cases in which air encephalograms and full neuropathological post-mortem studies increase this group will progressively diminish, many cases being transferred to Group 3.

(2) Cases where there is a demonstrable abnormality in the nervous system, but it does not affect the lower motor neurone, e.g. demyelinating and other diseases affecting the cerebellum.

(3) Cases where the lower motor neurone is affected but the lesion cannot be demonstrated in biopsies by classical methods. This group probably includes some of Walton's cases of benign congenital hypotonia, cases recovering after infantile polyneuritis, and possibly cases where the spinal cord has been injured by anoxia at birth as well as *formes frustes*, or extremely slowly progressive cases of Werdnig-Hoffmann disease.

The new techniques for demonstrating the intramuscular nerve endings are of the

greatest value in the third sub-group where they may be the only means of demonstrating during life involvement of the lower motor neurone. The nature of the changes, which are milder than those seen in classical Werdnig-Hoffmann disease, is described and the additional information already obtained by electron microscopy indicated.

RÉSUMÉ

Les bébés 'mous'

L'étude biopsique de prélèvements de terminaisons nerveuses intramusculaires chez 25 'enfants mous' a été faite par coloration vitale, mise en évidence histochimique de la cholinestérase et microscopie électronique. Les cas peuvent être répartis en deux groupes principaux:

(*a*) Ceux qui, à l'étude biopsique, par les méthodes classiques de coloration, présentent des aspects anormaux. L'anomalie le plus fréquemment rencontrée est l'atrophie bien connue des fibres musculaires par groupes indiquant la dégénérescence du neurone moteur inférieur. Cette dégénérescence, chez les 'enfants mous', est due en général à la maladie de Werdnig-Hoffmann. Dans cette maladie, les nouvelles méthodes d'étude révèlent un aspect très caractéristique des terminaisons nerveuses intramusculaires, d'une haute valeur diagnostique.

(*b*) Cas où, par les méthodes classiques de coloration, la biopsie musculaire est normale. Ceux-ci peuvent être, à leur tour, divisés en trois sous-groupes:

(1) Cas où ni l'encéphalogramme gazeux, ni l'examen neuropathologique complet après autopsie ne rélèvent d'anomalie du système nerveux. Ces cas semblent correspondre â la myotonie congénitale d'Oppenheim, quelques uns â l'hypotonie congénitale bénigne de Walton. Il est certain qu'â mesure que s'accroîtra la proportion des cas ox seront pratiqués l'encéphalogramme gazeux et un examen neuropathologique complet après autopsie, ce groupe s'amenuisera progressivement. De nombreux cas feront alors partie du groupe 3.

(2) Cas où il existe une anomalie du système nerveux mais qui n'affecte pas le neurone moteur inférieur, ex: démyélinisation et autres affections du cervelet.

(3) Cas où le neurone moteur inférieur est atteint mais où la lésion ne peut être décelée par les méthodes biopsiques classiques. Ce groupe comprend probablement certains cas d'hypotonie congénitale bénigne de Walton, des suites de polynévrites infantiles récupérables et peut-être des cas où la moelle épinière a été endommagée par une anoxie à la naissance, ainsi que les formes frustes ou des cas à évolution extrêmement lente de maladie de Werdnig-Hoffmann.

Les nouvelles techniques de mise en évidence des terminaisons nerveuses intramusculaires sont du plus grand intérêt dans le troisième sous-groupe où elles représentent le seul moyen de déceler, sur un sujet vivant, l'atteinte du neurone moteur inférieur. La nature des modifications, moins sévères que dans la maladie de Werdnig-Hoffmann, est discutée et les données supplémentaires, que révèlent déjà la microscopie électronique, sont indiquées.

ZUSAMMENFASSUNG

Bioptische Proben der Muskeln in der Diagnose von schlaffen Kindern.

Bioptische Proben intramuskulärer Nervenendigungen von 25 'schlaffen Kindern' sind mittels vitaler Färbung, histochemischer Prüfung auf Cholinesterase und elektronischer Mikroskopie untersucht worden. Man kann die Fälle in zwei Hauptgruppen aufteilen:

(*a*) Diejenigen, in denen die bioptischen Proben, mit klassischen Färbverfahren untersucht, abnorm erscheinen. Die am Häufigsten aufgefundene Abnormität ist die gut bekannte Atrophie der

Muskelfasern in Gruppen, die auf die Entartung des unteren motorischen Neurons hinweist. Diese Entartung ist bei den 'schlaffen Kindern' gewöhnlich durch die Werdnig-Hoffmannsche Krankheit verursacht. Bei dieser Krankheit zeigen die neuen Untersuchungsmethoden ein sehr charakteristisches Bild, von hohem diagnostischen Wert.

(*b*) Fälle, in denen nach Anwendung klassischer Färbmethoden die Muskelbiopsie normal erscheint. Diese kann man wieder in drei Untergruppen einteilen:

(1) Fälle in denen weder ein Pneumoencephalogramm noch eine vollständige neuropathologische Untersuchung post mortem irgend eine Abnormität des Nervensystems an den Tag bringen. Diese scheinen der Amyotonia Congenita Oppenheim's und einigen Fällen der gutartigen angeborenen Hypotonie von Walton zu entsprechen. Sicher wird diese Gruppe, mit der wachsenden Anzahl der Fälle in denen ein Pneumoencephalogramm und eine vollständige neuropathologische Untersuchung post mortem vorgenommen werden, nach und nach abnehmen. Man wird dann viele Fälle in die Gruppe 3 übertragen.

(2) Fälle mit nachweisbarer Abnormität des Nervensystems, die aber den unteren motorischen Neuron nicht berührt, z.B.: Demyelination oder andere Krankheiten des Kleinhirns.

(3) Fälle in denen der untere motorische Neuron angegriffen ist, in denen man aber die Schädigung mit klassischen Methoden auf den Biopsien nicht nachweisen kann. Diese Gruppe enthält wahrscheinlich einige Fälle von Walton's gutartiger angeborener Hypotonie, in Heilung übergehender infantile Polyneuritis und vielleicht Fälle, in denen das Rückenmark durch Anoxie während der Geburt geschädigt worden ist, sowie 'formes frustes' oder sehr langsam forschreitende Fälle der Werdnig-Hoffmannschen Krankheit.

Die neuen Untersuchungstechniken der intramuskulären Nervenendigungen sind von höchstem Wert in der dritten Untergruppe, in der sie das einzige Beweismittel, während des Lebens, für den Angriff des unteren motorischen Neurons darstellen. Das Wesen der Veränderungen, die ein milderes Bild als in der Werdnig-Hoffmannschen Krankheit aufweisen, wird beschrieben und die ergänzenden Befunde, die man schon durch elektronische Mikroskopie erhält, werden angegeben.

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FACTORS RENDERING THE CEREBRAL PALSIED CHILD CAPABLE OR INCAPABLE OF BENEFITING FROM FORMAL EDUCATION*

H. B. DAVIES, B.SC.(ECON.)
Principal, Thomas Delarue School, Tonbridge, Kent

To benefit from formal education the cerebral palsied child must have average intelligence and aptitude and good drive, accompanied by incentive, and adequate learning and physical aids. Because of the wide range of individual differences and his peculiar learning difficulties, his education must be largely individual.

A lowered capacity for intellectual activity in cerebral palsied children is the principal limiting factor to learning, but this is not indicative of their ultimate potential.

FORMAL education is concerned with the inculcation of the basic skills and fundamentals of learning which will enable an educable child to acquire facility in the use of language, both written and spoken, and in the use of number; with this he will be equipped to learn something of the world around him, of literature, history, the arts and sciences, in a normal school situation.

How far he will progress in learning depends on a number of factors, the most important of which is innate *intelligence*. It is essential, therefore, to have a sound assessment of the child's intelligence if we are not to risk forcing on the child a discipline for which he may be quite unsuited, and which may, in fact, do him serious harm. The connotation of this term intelligence may here include, as a corollary, aptitude or innate suitability.

Intelligence, however, must be accompanied by the will or desire to learn if satisfactory progress is to be made, and this will to learn is, unhappily, sometimes lacking in spastic children of good innate intelligence as they approach secondary-school age. It has been quenched out or damped down, perhaps by constant failure of achievement on the physical side; by being asked too much of at home or at school; by lack of opportunity; or by lack of stimulus or encouragement. This can happen, of course, with normal children as a result of ill-health, absence from school, frequent changes of school, and increasing failure to keep up with the class.

Experience during this last decade has clearly shown that cerebral palsied children of normal intelligence and drive, even when very severely disabled, and even with little or no speech, can cope with formal education provided they can receive individual attention in the basic subjects from enlightened and patient teachers, with the stimulus of group work during some periods and all in a secure, happy atmosphere. Many will need specially designed or adapted furniture, specially written and designed books, and a variety of learning aids, and individual teachers and therapists have shown remarkable ingenuity and versatility in designing these aids and furnishings. There has been a regrettable failure on the part of commercial educational and medical suppliers to do much to help in this connection, though they have done more to meet the physical needs of

these children than their peculiar learning needs.

Concurrently with the child's formal school work must be the treatment of his physical handicaps, despite the bogey that these treatment sessions often present to the striving teachers and timetable planners. The pupil must be allowed time to readjust himself to the classroom situation on returning from therapy, and also to ensure as full a carry-over as possible of the therapist's work into the classroom. This entails the fullest possible understanding between teacher and therapist if the work of either or both is not to be jeopardised to the detriment of the pupil. With great respect I would suggest that a better understanding of and sympathy for the work of the teacher by some of our medical advisers would alleviate and help to remove the friction and clash of interests which sometimes develop between teachers and therapists. Therapy is the ancillary of education in the schools, just as education is often the ancillary of treatment in the hospitals.

The intelligent cerebral palsied child, eager to learn, needs and should derive great encouragement and incentive from his therapists and doctors if he is not to chafe and fret over his physical handicap in the classroom. Given, therefore, intelligence, drive, incentive, and the necessary learning and physical aid there is ample proof that he can benefit from formal education.

Need for Individualisation

This formal education must, however, be individual to a very large extent, because there are even more individual differences among cerebral palsied children than among normal ones. Apart from differences of age, ability, aptitude and attainment there are the whole range of physical disabilities and the peculiar learning difficulties. It is a moot point how far all these learning difficulties are peculiar to cerebral palsied children, but certainly some appear to be unique. We have found, for instance, that pupils who have difficulties in spatial perception appear to have lapses of memory relating to a particular arithmetical process, notably in subtraction and division, less frequently in multiplication, and *never* in addition. At such times the pupil seems incapable of reasoning on that particular process yet is capable of logical thought on another branch of the subject. Next day he often can remember and use the process previously forgotten. There appears to be a direct relation between the degree of lack of spatial perception and the intensity of these mental 'black-outs'. During these lapses some of these children roll their eyes upwards and appear to derive physical pleasure from so doing.

Some highly intelligent pupils with good hands, able to write easily and quickly, have manifested another curious inhibition of memory. They will answer oral questions well, revealing a sound knowledge of advanced-level subjects, but when required to write the same questions fail dismally. Yet when four such pupils were allowed to dictate their answers to an amanuensis, one passed General Certificate of Education in mathematics with distinction, one passed with distinction in both mathematics and French, one passed with distinction in English, and the fourth passed with distinction in French! All showed a degree of distractability. This seems to emphasise the tremendous significance of the psychosomatic factor in cerebral palsy.

Why do some pupils, who are only lightly physically handicapped and able to write easily, cope well with mathematics and yet reveal abnormal difficulty in the English subjects? These same children all show a greater or lesser degree of distractability. May it be that because mathematical problems have only one answer, the mind when it returns to such a problem must ultimately return to the previous line of reasoning, whereas in an essay a line of thought once lost may well be lost for ever?

An interesting and amusing example of perseveration occurred at a party on Hallow E'en. A boy had joined in an apple-bobbing competition, but the biting action made such an impression on him

that some 20 minutes later he attempted to bite the light from a torch shining behind a sheet, which he was asked to follow with his nose!

Learning Difficulties

A number of specific learning difficulties are common to cerebral palsied and to educationally subnormal (ESN) children, such as shortness of visual perception, including span of perception and span of recognition, difficulties in the concept of number, distractability, perseveration, lack of drive, faulty teaching, difficulties due to loss of experience through environment or disability, over- or under-dependence. It is interesting to speculate on whether some categories of the ESN are brain-injured children, and conversely whether some of the learning difficulties of the cerebral palsied child are caused by emotional disturbances.

A lowered capacity for intellectual activity is perhaps the greatest limiting factor to learning and benefiting from formal education, apart from the purely motor manifestations in the cerebral palsied child. We must accept the fact that a greater proportion of these children function at a lower level than children in the ordinary population, but this does not indicate what their potential level may or will be. New trends in treatment seem to hold much promise even for those whose capacity for voluntary control is poor. Every cerebral palsied child should be regarded as educable until proved otherwise, and this makes experienced assessment panels and assessment centres absolutely essential.

Formal education is not for the brain-injured child whose intelligence is of a low order, and here we fall back on informal education. This may include socialisation, sensory training and the specialised methods of the special schools for ESN children which need not concern us here.

Medical and Educational Advice for Teachers

To come now to the help which teachers would welcome on the medical and educational aspects of the care of cerebral palsied children. From our medical advisers we need an increased emphasis on the effects of cerebral palsy on the child's learning potential, on the inter-relation between the physical and the mental impairment. We need a closer liaison between medicine and education. We would welcome more advice and instruction through courses, papers and discussions, for we must have adequate understanding of the mental, physical and emotional patterns characteristic of the brain-injured.

We would like to see our doctors visit us in the classrooms sometimes, to see our problems in action and to see their patients in the classroom situation. Such visits would be very reassuring.

We would like our educational advisers to consider and seek solutions to the peculiar learning difficulties of the cerebrally palsied; to consider, examine and suggest learning aids, furniture and equipment; to review and recommend textbooks for teachers and pupils and to review specialist publications. We would like them to consider and make recommendations about adult and further education, and about vocational training courses for the brain-injured; to review the need for specialised training courses and for some correlative theoretical instruction and general education for trainees.

Recommendations about recreative and leisure-time activities for spastics at home or at centre would be appreciated. How can we help the severely handicapped to maintain an interest in life?

And what of the special problems of the *ineducable spastic*, his training and management, and of the severely handicapped, intelligent and maybe well-educated, but unemployable spastic?

Recording Experience

There is undoubtedly a great body of expertise to be found in the schools and centres. Ideally teachers and therapists should record carefully their successes and their failures with each individual child, the means by which at each stage they

overcome learning or motor difficulties, the special tricks, improvisations, modifications etc., used, and with what effect. But alas, they are busy and preoccupied all day and exhausted at the end of it, and so these precious records are rarely written up. This knowledge must be collected and made available to all. Teachers and therapists are for ever designing, modifying and adapting furniture and apparatus for individual pupils, but no drawings or records exist in most cases. We need to know what special gadgets are proving valuable as aids to learning; what special furniture is proving helpful; what size and kind of type, what kind and shape of book are most useful at the various stages of learning to the spastic with disabled hands, defective vision, or so on.

The Medical and Educational Advisory Committees and the Assessment Panels should together devise a standard form covering the pre-school and school history of every C.P. child. This should include information about home conditions, parental attitude, physical and mental achievements to date, home environment related to learning opportunities, encouragement to independance, follow-up of therapy and learning. It should include information regarding the development of speech and other means of communication and learning—for example, whether he can chew, suck, blow, point, nod, etc., and his responses to various stimuli, such as colour, smell, taste, movement, rhythm, music and non-musical sounds, objects animate and inanimate around him, whether he can sort colours, shapes, sizes; whether he can count except purely by note. If he reads, what level is he at? What is the cleverest thing he can do at home? What does he most like doing, and so on. The vital question is, of course: '*Can we teach this child?*'

There is a great need for full, accurate and long-term records of C.P. children, records that can be compared from year to year in a concrete and measurable manner. Unobtrusive photography and tape-recording could add much to the value of such records. With a standardised record like the one proposed, and with the annual performance of the Binet Scale as a check on the efficacy of treatment and education in improving overall facility of expression, it should be possible in a few years' time to use such data in formulating a more adequate and scientific method of evaluation and prognosis.

SUMMARY

Innate intelligence, which includes aptitude, is the principal factor in determining a child's learning ability. It must be accompanied by the will to learn, and this is often lacking in spastic children through constant failure on the physical side, lack of opportunity, and by lack of stimulus or encouragement.

Given intelligence, drive, incentive and adequate learning and physical aids, the intelligent cerebrally palsied child can benefit from formal education. In view of the individual differences in age, ability, aptitude, attainment, and the whole range of physical disabilities, allied to his peculiar learning difficulties, his education must be largely individual.

Certain serious learning difficulties are common to cerebrally palsied and educationally subnormal children, including shortness of visual perception, span of perception and recognition, difficulties in the concept of number, distractability, perseveration, lack of drive and loss of experience. Are some categories of educationally subnormal children brain-injured? And are some of the learning difficulties of cerebrally palsied children caused by emotional disturbances?

The greatest limiting factor to learning is a lowered capacity for intellectual activity, and cerebrally palsied children generally function at a lower level than ordinary ones, but this must not be taken to indicate their potential level of achievement. Every cerebrally palsied child should be regarded as educable until proved otherwise, if necessary at an Assessment Centre.

The brain-injured child of low intelli-

gence requires informal education, including socialisation, sensory training and the specialised methods of the schools for educationally subnormal children.

Full, accurate and long-term records, comparable from year to year in a measurable manner, are urgently needed for the formulation of a more adequate and scientific evaluation and prognosis of the brain-injured.

RÉSUMÉ

Enfants infirmes moteurs cérébraux. Facteurs les rendant aptes ou non à tirer parti d'un enseignement traditionnel.

L'intelligence innée, qui comprend l'aptitude, est le facteur principal dans la détermination de la capacité d'apprentissage d'un enfant. Elle doit être accompagnée de la volonté d'apprendre qui fait souvent défaut aux enfants spastiques en raison de constantes défaillances physiques, du manque d'opportunité et aussi du manque de stimulus ou d'encouragement.

S'il est doté d'intelligence, d'émulation, d'un matériel éducatif et physique adéquat, l'enfant intelligent atteint d'infirmité motrice peut profiter d'un enseignement traditionnel. Mais, étant donné les différences individuelles d'âge, de facultés, d'aptitude, de résultats obtenus, auxquelles viennent s'ajouter ses difficultés particulières d'apprentissage, son éducation doit être nettement individuelle.

Les enfants atteints d'infirmité motrice cérébrale et les enfants retardés scolaires ont en commun certaines difficultés spécifiques d'apprentissage: perception visuelle courte, perception et recognition limitées, conception des nombres difficultueuse, distraction, idées fixes, manque d'allant et perte d'expérience. On se demande si certaines catégories d'enfants retardés dans leurs études à la suite d'une lésion cérébrale et certaines difficultés d'apprentissage des enfants infirmes moteurs cérébraux ne proviennent pas de troubles affectifs.

Le plus grand facteur limitatif de l'apprentissage est une capacité réduite de l'activité intellectuelle; or les enfants infirmes moteurs cérébraux fonctionnent généralement sur un registre plus bas que les enfants ordinaires, ce qui ne préjuge en rien de la réussite dont ils sont virtuellement capables. Tout enfant atteint d'infirmité motrice devrait être considéré comme éducable jusqu'à preuve du contraire, faite au besoin dans un centre d'orientation.

Il convient de donner à l'enfant infirme moteur cérébral de faible intelligence une éducation non traditionnelle, comprenant la socialisation, l'éducation sensorielle et les méthodes spécialisées des écoles d'enfants retardés scolaires.

Des dossiers complets, précis et à long terme, susceptibles d'être mesurés comparativement d'année en année, sont d'une nécessité urgente pour la mise au point d'une évaluation et d'un pronostic plus adéquats et plus scientifiques des cas d'infirmes moteurs cérébraux.

ZUSAMMENFASSUNG

Faktoren, die Kinder mit zerebraler Kinderlähmung fähig oder unfähig machen, Nutzen aus der üblichen Ausbildung zu ziehen.

Die angeborene Intelligenz, die die Begabung einschliesst, ist der hauptsächlichste Faktor für die Bestimmung der Lernfähigkeit eines Kindes. Sie muss von der Lernbegierde, die oft bei den spastischen Kindern wegen beständiger körperlichen Schwächen und aus Mangel an günstigen Gelegenheiten, an Antrieb und Ermunterung, fehlt, begleitet sein.

Mit Intelligenz, Antrieb und angemessener pädagogischer und physikalischer Hilfe kann ein intelligentes Kind mit Zerebrallähmung aus dem üblichen Unterricht Gewinn ziehen. Aber in Hinblick auf die individuellen Unterschiede des Alters, der Begabung, der Fähigkeiten, der Erreichungen und aller körperlichen Hemmungen, verbunden mit seinen eigenen Lernschwierigkeiten, muss seine Erziehung zum grossen Teil individuell sein.

Gewisse spezifische Lernschwierigkeiten findet man sowohl bei Kindern mit Zerebrallähmung als auch bei Kindern mit

Intelligenzdefekt auf: zu kurze optische Wahrnehmungen, beschränktes Wahrnemungs-und Erkennungsvermögen, Störungen des Zahlenbegriffs, Ablenkbarkeit, Perseveration, Antriebsmangel und Verlust der Erfahrung. Haben gewisse Kategorien schwachbegabter Kinder Zerebrallähmung und sind gewisse Lernschwierigkeiten der Kinder mit Zerebrallähmung durch emotionale Störungen verursacht?

Der Faktor, der das Lernen am meisten beschränkt, ist eine verminderte Leistungsfähigkeit der intellektuellen Tätigkeit. Bei den Kindern mit Zerebrallähmung steht diese meistens niedriger als bei den gewöhnlichen Kindern, aber man darf nicht daraus auf ihre endgültige Möglichkeiten schliessen. Jedes Kind mit Zerebrallähmung sollte als erziehbar betrachtet werden bis zum Beweis des Gegenteils, nötigenfalls in einer Beratungsstelle.

Für das Kind mit Zerebrallähmung und Intelligenzdefekt ist eine spezielle Erziehung, die Förderung der Geselligkeit, sensorische Trainierung und die besonderen Methoden der Anstalten für Schwachbegabte verknüpft, angezeigt.

Vollständige, genaue und lange durchgeführte Krankenprotokolle, die von Jahr zu Jahr auf messbare Weise vergleichbar wären, sind für die Formulierung einer angemesseneren und wissenschaftlicheren Bewertung und Prognose der Fälle von Zerebrallähmung drigend nötig.

EDITORIALS

WRITINGS OF HUGHLINGS JACKSON*

'We must remember that many doctrines were stated years ago in principle which were then novel and much disputed, but are now so generally accepted that we are in danger of ceasing to think of the very early propounders of those doctrines.'

THESE words, with which John Hughlings Jackson modestly disclaimed originality for his own work, might appropriately have been used today in referring to Jackson himself, for the principles he so clearly enunciated nearly a century ago are the fabric of modern neurology. Jackson never wrote a textbook of neurology, and much of his work might have remained buried in the hospital reports and medical journals of his time had not the guarantors of *Brain* decided to republish some of his papers. We owe a great debt to them and to Dr. James Taylor, who selected from Jackson's voluminous writings the most important and representative of his articles for publication in two volumes of collected writings. These were first published in 1931 but have long been out of print, and their reappearance from America is therefore most welcome. The new publication is an exact replica of the first edition, even down to the misprints.

The first volume is a selection of Jackson's papers on epilepsy and convulsions and includes among others his classical descriptions of local epileptiform seizures—the Jacksonian fits by which his name is familiar to every medical student. Notwithstanding their importance, these represent only a part of the wealth of information about epilepsy contained in the volume. His penetrating studies of convulsive seizures led directly to his generalisation that the Rolandic area of the brain is motor, and that the movement of each part of the body is represented there. Although some of his other hypotheses are outdated and we may weary of his long exposition on venosity of the blood as a cause of 'inward fits' (laryngismus stridulus), Jackson's meticulous observations and detailed descriptions of clinical phenomena remain as valid today as ever they were. His views on the localisation of cerebral movements, on dreamy states and on postepileptic states changed the face of neurology, and alone entitle Hughlings Jackson to a pre-eminent place among the pioneers in this field.

The second volume contains papers, addresses and lectures on the evolution and dissolution of the nervous system, on affections of speech and on a variety of other subjects. Because of his insistence on the precise use of words and the pains which he always took to avoid misrepresentation of his views, Jackson's writing is at times tendentious and often repetitious, but he seldom fails to hold the reader's interest. Whether he is pleading eloquently for the wider use of

* *Selected Writings of John Hughlings Jackson.* Edited by James Taylor, with the assistance of Gordon Holmes and F. M. R. Walshe. New York: Basic Books Inc., 1958. 2 vols., pp. 500 and 510. $15.

the ophthalmoscope in clinical medicine or insisting on the necessity for recording the minutest details of a convulsion, his words have as clear and pertinent a message for physicians today as when he wrote them. Jackson's work is liberally sprinkled with pithy aphorisms on the wider aspects of medicine such as, 'If a man can take care of his arteries . . . his nervous tissues will take care of themselves' and, of specialisation, 'There is no harm in studying a special subject: the harm is in doing any kind of work with a narrow aim and with a narrow mind'. At times he shows his impatience with the authoritarian dogmatism of contemporary medical thought, as when he says, 'We have long heard that old maids' husbands are always well behaved, and on the same principle the pathology of those who do not make post-mortem examinations is often confident and definite'.

These volumes of writings cannot fail to interest those working in the field of cerebral palsy. They will find here much that they had believed to be newly discovered during the feverish activity of the past few years. The value of Jackson's work is not diminished by his limited knowledge of the extrapyramidal control of movements, and his remarks often have a startlingly modern ring. Such observations as 'Nervous centres represent movements, not muscles. From negative lesions of motor centres there is not paralysis of muscles, but loss of movements', and 'We ought . . . to distinguish most carefully abnormal functional changes from the pathological processes leading to them' might have been taken straight from a recent text on cerebral palsy. Although we may not agree that 'rigidity (in hemiplegia) is owing to unantagonised influence of the cerebellum', other statements, such as that 'tremor . . . is a minor degree of tonic spasm', force us to make a fresh appraisal of our concepts of motor dysfunction. Most of Jackson's work is based on experience with adults, but his observations on such conditions as breath-holding and head-banging are of great paediatric interest, and when he says 'we may narrow a young epileptic girl's life too much by forbidding the amusements proper to her age' we are again sharply reminded how far he was ahead of his time.

In these two volumes we can follow the development of Hughlings Jackson's thought from his careful observations in the case-reports of the 'sixties, through the formative years of the 'seventies, when he marshalled his ideas with increasing precision, to the full flowering of his views in the Croonian Lectures on the *Evolution and Dissolution of the Nervous System* (1884) and the Lumleian Lectures on *Convulsive Seizures* (1890). Many of his theories may have been modified with the passage of the years, but the principles which he set forth have stood the test of time, and we can only marvel at the power of his intellect and the lucidity of his thought.

Sir William Osler said of Hughlings Jackson barely a month before he died that 'some of the most deserving men never receive any recognition'. What better recognition of his contribution to medicine could be made than the republication of his collected writings nearly fifty years after his death?

R. G. MITCHELL

RESPIRATORY DISTRESS SYNDROME OF THE NEWBORN

THE most urgent problem facing the newborn infant is the establishment of pulmonary respiration. If the infant fails he will die; if he succeeds only partially he will endure a period of anoxaemia with possible irreversible damage to brain cells. Himwich and her colleagues[5] have recently attempted some metabolic studies on perinatal human brain which show the great need for oxygen of the cerebral cortex, the thalamus and the basal nuclei.

The very immature infant may fail to initiate pulmonary respiration and at autopsy primary atelectasis and anoxic cerebral haemorrhage is the most frequent finding. More puzzling are those infants who develop their respiratory distress shortly after birth and display what used to be termed 'hyaline membrane disease'. This syndrome has recently been well reviewed by James[6].

The morbid anatomists have established some basic facts about this interesting condition. The essential features are the bulky, congested lungs which on section show varying degrees of atelectasis and the presence of eosinophilic membranes lining the terminal bronchioles and alveoli. The membrane contains fibrin and is likely to be endogenously produced. The membranes are never found in infants who have not breathed and the appearance is confined to premature infants, those born by Caesarean section, and infants of diabetic mothers. The clinical course and radiographic appearances are fairly characteristic.

The aetiology is unknown, so that treatment is empirical and unspecific. Reliance is placed on good nursing and the administration of oxygen, while the infant is left unencumbered to make the best use of his respiratory efforts. The search for a cause has been unrelenting but so far unrewarding. The early postulate that this was an inhalation phenomenon is unlikely. Many ingenious haemodynamic explanations based on the circulatory adjustments of neonatal life did not fully explain the known clinical or pathological facts. Recently opinion is reverting to consideration of the lung once more as the primary source of the disorder. Clements and his co-workers[2] have been interested in the surface tension of the lining of the lungs. They have found evidence of a substance, probably a mucoprotein[3], which has the property of lowering the alveolar surface tension. They point out that a low surface compressibility within the lung means that there is an increased range of stability for the individual alveolus and also that the number of alveolar sizes which can coexist in the lungs at any given pressure is increased.

Such advantages are necessities for the newborn lung. With some alveoli opening while others remain uninflated it is obviously important for those expanded to remain open, and it is equally necessary that further inspired air does not over-expand the already inflated alveoli while others remain atelectatic. The surface-tension-lowering

agent then protects the lungs against both emphysema and atelectasis.

Deficiency of such a substance would not necessarily hinder primary expansion because the problem here is breaking the fluid film which holds the alveolar walls together. It would, however, cause grave subsequent difficulties. The alveoli would tend to collapse at each expiration and could be held open only by strong intrathoracic negative pressures. These have been demonstrated by Karlberg *et al.*[7] in infants with respiratory distress. Such intense negative pressures could suck out fluid from the pulmonary capillaries, creating the 'hyaline membrane', and also disturb the filling of the heart, leading to congestive failure.

This hypothesis is plausible, and if it could be shown that immature infants lacked the necessary surface-tension-reducing substance it would be almost convincing. Such a substance would have no utility in foetal life, so there is no reason for its presence in the foetus. Avery and Mead[1] have shown that the surface tension of the washings of lungs from immature infants and infants dying of respiratory distress is increased. Gruenwald[4] has briefly reported a series of experiments showing that some, but not all, stillborn premature infants have poor stability of lung expansion. His work is supported by the American United Cerebral Palsy Research and Educational Foundation.

It is tempting to relate hyaline membrane to yet another physiological immaturity of the premature infant. This leaves unexplained the susceptibility of the infant born by Caesarean section. Much further work is needed, but this seems the most profitable pathway to explore.

T. E. OPPÉ

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NEW LIGHT ON THE PLANTAR REFLEX

NEARLY half the issue of *Brain* for December 1959 (vol. **82,** part 3) is taken up with two papers on the plantar reflex. In the first, Sir Russell Brain and Dr. M. Wilkinson describe their observations on 35 normal infants aged from 1 day to 2¼ years; on 31 assorted cases of cerebral palsy in children; and on five adults with long-standing lesions of the spinal cord. In each group they stimulated the sole, thigh, abdomen (T 10), chest (T 3) and neck on both sides, with pin-prick, cold and, in the case of the sole, 'key-drag'. They usefully set out their full 2800-odd observations on the ensuing behaviour of the great toe in one large page of tables. This clearly shows a pattern of change in the maturing nervous system of the normal infant. From 1 to 8 days the response can be elicited by pin-prick or cold from all levels up to T 3; it is always extensor, but usually ipsilateral only. From 5 to 20 weeks when elicited from the sole and thigh the response is always extensor and usually bilateral; from the abdomen and upwards it is equally often flexor or extensor and usually bilateral. After 21 weeks the receptive field shrinks from above downwards, and cold ceases to be an effective stimulus after 24 weeks. After 32 weeks bilateral responses become rarer, and the doubtful or frank flexor response becomes progressively commoner, although (unfortunately) it is by no means established as invariable by the time their normal group ends at 2¼ years. There is, as others have pointed out, a suggestive parallel time-course between these reflex changes and the deposition of myelin in the cortico-spinal tract, but our present knowledge does not enable us to assert any causal connection.

However, the state of affairs found in normal infants is not found in children or adults with lesions of the central nervous system. The response in the cerebral palsied children and in the adults with spinal cord lesions showed no evidence of the widespread receptive zone found in infants, and the authors suggest that in the infant this reaction is perhaps partly a general reaction to cold which serves a homeostatic purpose until the temperature-regulating mechanism is fully established. They have further studied the associated movements of the great toe in 30 normals and 30 patients with 'a pyramidal lesion', and they observed that extension occurred in a third of the normals in association with flexion of the knee and hips against resistance. In the abnormal group the same manoeuvre elicited extension in two-thirds. It thus appears that a lesion can modify an associated movement, and Russell Brain and Wilkinson conclude that 'the difference between the pathological flexion reflex of the lower limb, of which the extensor plantar is part, the normal flexor plantar reflex and a voluntary movement of flexion is not a difference of kind but of the nature and degree of physiological organisation exhibited'. The reader might wonder how motor reflex patterns could differ except in the nature of the physiological organisation, but the second paper by Dr. W. M. Landau and Dr. M. H. Clare sheds valuable light on this important point of detail.

Here an electromyographic and film technique has been used to analyse the behaviour of individual muscles during the flexor and extensor types of plantar reflex, as shown in 14 normal people and 20 patients with various acute and chronic lesions. The main conclusions reached by Landau and Clare is that 'the extensor reflex is not a different reaction from the flexor but rather a hyperactive flexor response in which the extensor hallucis longus is included by radiation'. The outward effect of the inclusion of the long extensor is that the action of the weaker flexor group is overridden and the great toe extends. Apart from film and electromyographic evidence for this they have shown that one can convert an extensor response into a flexor by a peroneal nerve block, and that when an extensor response paradoxically fails to appear in cases of amyotrophic lateral sclerosis this may be a result of atrophy in the long extensor muscle. The evidence that the 'flexor' and 'extensor' responses are not independent muscular synergies but qualitative and spatial variations of the same pattern is further strengthened by the features they are shown to have in common—minimal thresholds on the side of the foot; and a motor focus of contraction in the flexor muscle of the hallux, ankle, knee and hip.

Taken together these two papers usefully clear the air. The idea that a new reflex appears with disease or functional disturbance of the central nervous system was always unattractive to the physiologist and is now superfluous. The validity or utility of 'Babinski's sign' is unimpaired. This sign has little value for predicting microscopic changes in the cortico-spinal fibres, but it will always remain extremely valuable for directing the clinician to some change in the functional state of the central nervous system, for which he may then find a cause.

J. A. V. BATES

AN OPERATION FOR PARALYTIC HIP CONTRACTURE

SOME good results have been obtained at the Makerere College Medical School, in Uganda, with a simple procedure for the correction of hip flexion contracture following poliomyelitis. The operation was developed by Robert Roaf in India, and he demonstrated it in Uganda in February, 1958. It is based on the assumption that the contracture which follows paralysis of the hip extensors is largely a result of shortening of the ileo-tibial tract, which produces a flexion contracture not only of the hip but also of the knee. The tract, together with its deep attachments to the femur, is divided subcutaneously at several points along the lateral aspect of the thigh and hip. The shortened ileo-psoas muscle and the joint capsule are then gradually stretched as the hip and knee are extended. A plaster cylinder is then applied to the leg, and walking may be allowed in a few days. When the contracture is severe a hip spica may be required.

David Allbrook and H. Fletcher Lunn (*Lancet*, 1960, **i**, 459) have operated on 32 children in this manner, and over a period of 18 months there was no evidence of recurrence of the deformity. Flexion contracture of the knee was corrected with ease, and posterior capsulotomy was not required.

The chief indication for this procedure is in deformities following poliomyelitis, but it may also have a place in the correction of hip deformities arising in cerebral palsy.

J. S. Batchelor

REPORT

SENSORY, AUDITORY AND SPEECH DEFECTS IN CEREBRAL PALSY

A DISCUSSION ARRANGED BY THE MEDICAL ADVISORY COMMITTEE OF THE N.S.S. FOR THE HEADS AND THERAPISTS OF THE SOCIETY'S SCHOOLS AND CENTRES

This discussion was held at the Hospital for Sick Children, Great Ormond Street, London, on May 9, 1959, with Dr. J. H. Crosland in the Chair. The main speakers were Dr. J. P. M. Tizard, Dr. L. Fisch, and Mrs. D. E. Back.

Sensory Defects

DR. J. P. M. TIZARD, of the Institute of Child Health, London, said that little attention has been paid to sensory defects in cerebral palsy until recent years.

Most children, he said, are first brought for examination at an age when it is not possible to undertake sensory testing, and medical students are often not told the importance of carrying out further examinations after the initial one. This talk would be mainly concerned with hemiplegia. In most cases of hemiplegia the cerebral defect is perinatal in origin, while in the remainder some obvious and dramatic postnatal incident in the first few years of life has led to the first signs of hemiplegia. Over half of infantile hemiplegics have sensory defects. As a rule the sensory defects take the form of some loss of peripheral limb sensation, but there are visual-field defects in about a quarter of these children. It is very unusual to have a field defect without any sensory defect in the affected limbs. The sensory loss does not usually involve simple sensations such as touch, pain, temperature, etc. If the thalamus is intact these forms of sensation will be preserved. It is powers of discriminatory sensation, appreciated at a cortical level, that are affected—i.e., the ability to appreciate shape and texture, the position and movement of joints, and the ability to differentiate between touch of one or two points. Sometimes simple forms of thalamic sensation are lost, usually in the postnatal hemiplegic group. Otherwise, there is no difference in the type or degree of impairment in the two groups.

It is much easier to establish with confidence that a young child is unable to appreciate one type of sensation, when normal ability to appreciate this in a normal limb can be demonstrated, than it is to be sure of the existence of sensory defects in bilaterally affected children. It is thus easier to detect sensory defects in cases of hemiplegia where one has the normal side to compare with the affected one, than it is to determine whether or not sensory defects exist in cases of quadriplegia. Moreover, quadriplegics often have defective intelligence: one might expect to find sensory defects in some 'double hemiplegics' if this term can be said to represent a valid subdivision of quadriplegia.

Methods of Testing

Both children and adults do their best to 'cheat' in tests. This is interesting and annoying and one must try to eliminate it. The true significance of the cheating is that

it is the method by which the child or adult manages in ordinary life. Many children and adults dislike being shown that they have these sensory defects.

Dr. Tizard then brought in Ursula, a girl of 12 who was normal until she was knocked down by a car at the age of six. She was unconscious and was operated on for a depressed fracture of the skull. In 1957 she was brought to Dr. Tizard because her right hand and arm were not growing normally. She had always been left-handed, even before the accident, but now hardly used her right hand at all, although there was little loss of power and practically no spasticity. Ursula's right arm was 1 inch shorter and her right hand ½ inch shorter than the left. Dr. Tizard demonstrated that Ursula had no loss of touch but had lost discriminatory sensation in three fingers of her right hand. He pointed out that patients tend to alternate their answers and this must be taken into account when testing. When tested for stereognosis, Ursula could not tell what she was holding in her right hand, though she could easily name the same objects—a key and various coins—when she held them in her left. Tests for feeling and other sensations should be allied to the patient's intellectual and cultural level; one must also make sure that the inability to appreciate shape and texture is not due to inability to manipulate. From the patient's point of view this amounts to the same thing, but not from the scientific point of view. In testing for visual-field defects it is better to use the confrontation method, because of 'cheating', and to use two stimuli instead of one.

Dr. Tizard discussed the extinction phenomenon, in which a patient can detect a single stimulus on the affected side, but fails to do so if the same stimulus is applied simultaneously to the normal side. He then showed a film demonstrating the half-field visual defect.

Speaking on the under-growth of limbs, Dr. Tizard said that when there is under-growth there is often a sensory defect as well, and it is unusual to find sensory defects without under-growth of the affected limb. This is not only muscle wasting but real shortening, the bones not having grown normally. Under-growth does not bear any close relationship to motor defect. If there is shortening of the limb this should alert one to the possibility of a sensory defect. Sensory defects bear no relationship to the time of onset of hemiplegia. There is some relationship to the degree of motor impairment but none to convulsions or intelligence.

Effects of Sensory Loss

Discussing the practical significance of sensory loss in hemiplegia, Dr. Tizard said that the learning of skilled voluntary movement depends on the ability to store up memories of previous movements made correctly and incorrectly, and a child with peripheral defects may not be able to do this. Training must comprise visual attention to movement. Half-field vision does not appear to matter much to the patient, except in driving a car.

QUESTIONS

Dr. Tizard asked the therapists what they felt about means of improving the usefulness in an arm when a child had a sensory defect such as Ursula's. Such a child could not accurately know the position of the affected limb in space. The therapists could not think of an answer to the problem. A blind child with bilateral sensory defects of this type would probably learn nothing.

Mr. Davies asked if incomplete body image was connected with this problem and Dr. Tizard agreed that the limb would be ignored. The children must be taught to watch all the time, as had happened with a hemiplegic boy at the Thomas Delarue School, who had learnt to use a plane with two hands and now used both his hands when eating.

Hearing Defects

DR. FISCH, research otologist at the Royal National Throat and Ear Hospital,

began by talking about problems of hearing in C.P. children with no impairment of hearing. The fundamental function of hearing, he said, is direction-finding. Then visual aids are added for judging distance. Head movements are also important for this and if one cannot move one's head at will one cannot use this combination of learning. Listening is an important function of hearing; one tries to adopt the best position of the body and head and to eliminate one's own noises. This may be difficult for a C.P. child to achieve. A C.P. child will find difficulty in obtaining information from its environment by the combination of these two types of communication. Some sources of sound may puzzle a C.P. child for longer than a normal child. A C.P. child cannot stop and listen. His optimum listening conditions may be bad, because of the movement of his body and his chair. Dr. Fisch was referring mainly to athetoid children who found it difficult to keep still and listen.

Effects of High-frequency Deafness

Some words may be lost or confusing to a deaf child and lip-reading is important to C.P. children. Even with a hearing-aid they may not be able to learn unless they can see the teacher and read his lips. They should be trained to look at the person talking to them. They should be helped to control their head better and the source of sound should be brought into their visual field, if necessary.

Hearing-aids

Proper training should be given when a child is provided with a hearing-aid. There are many difficulties in the use of an aid in the more severely affected C.P. child. Often there are technical difficulties regarding the size of the hearing-aid and the cords attached to it. It is now possible to obtain an aid fitted to the head and there are aids for both ears, to restore stereophonic hearing; these may be a little more expensive but are much more efficient. Rubbing noises from the body are unpleasant to the child when an aid is worn on the body, and this is the reason for advocating aids worn on the head.

The C.P. child often has a high-frequency deafness, without his low-frequency hearing being affected. Amplification of low-frequency sounds is unpleasant for these children and may make them reject the aids altogether. It is possible now to provide hearing-aids in which the low-frequency sounds are lowered.

Auditory Training

This consists principally of teaching the child to attach the correct meaning to the speech sounds which he can hear. He may not hear the full acoustic shape of speech and it may be a different sound that he hears. It is as if he learns a new code. Auditory training should not be confused with speech training.

More emphasis should be placed on the communication difficulties of the C.P. child, which may be very great when he is also partly deaf. A child may be able to understand much more than he can speak and should not be judged on his inability to speak intelligently. Understanding is often greater than executive speech itself. The first task is to achieve understanding.

Testing the Hearing of C.P. Children

There is great difficulty in the early detection and testing of very small children for deafness. The tests cannot be completed at a single examination. A child often has to be trained to perform the testing procedures, and this may take several weeks or even months. At the Centre for Spastic Children, Cheyne Walk, all the children are tested for hearing defects. Dr. Fisch hoped that the testing methods would be improved. They are trying to devise objective tests of hearing for C.P. children, so that the active co-operation of the child is not needed. Not enough work has been done on training people to apply the tests. Many years are often lost when children are put into mental institutions and are later proved educable but deaf. In spite of publicity and talks to the public, children are still found

in these institutions as a result of a mistaken diagnosis.

QUESTIONS

Asked how often the tests should be repeated, Dr. Fisch replied that there was no necessity to do the tests frequently, provided the first test was considered reliable, as the hearing would not change. It was important to repeat the test if it was inconclusive. The hearing defect in athetoids, which was the result of permanent damage, did not change. If motor control improved and auditory training was given, better use would be made of the hearing, and the ability to communicate would improve. If there was a discrepancy between the results of various tests it was because one or all the tests were inaccurate and further testing would be necessary until a consistent and reliable result was obtained. Training in carrying out tests was important because reliability of response could only be judged by an experienced person.

Dr. Fisch was asked if a child who lip-read before having a hearing-aid should be prevented from lip-reading when given an aid. He said that in formal lessons one could eliminate lip-reading and help the child to discriminate speech sounds by hearing alone. It was best for the child to use his hearing-aid and lip-reading together for communication.

It was suggested that lip-reading was not useful for communicating with a stranger. Dr. Fisch thought this was more likely when, in deaf schools, pupils were taught by exaggerated movements of the lips. The child might lip-read what the teachers said but might not understand strangers. One should talk to the deaf child normally, without exaggerated lip movement and forced articulation.

Speech Therapy

MRS. BACK, speech therapist at the Cheyne Walk Centre, said that 50 to 75 per cent of children with cerebral palsy will have some disorder of speech ranging from complete absence of language to a minor rhythmic disruption. The speech therapist should aim for useful speech, not perfect articulation. She must help them to express themselves, thereby helping them to attain a more pleasing personality.

Mrs. Back is now working with children ranging in age from two to seven years, but she has also worked with the school-age group and adults. All three groups must have an entirely different approach, but underlying all treatment is not only the child but his family, who are concerned in all stages of treatment and require guidance and real practical help. The teacher is an important ally because, in school, language and its purpose become a dynamic force in the child's life.

Cerebral palsied children may have any of the following difficulties: language disturbances, dysarthria, voice and breathing disorders, and hearing loss. Failure to develop speech and language may be due to a variety of causes, including difficulties of perception, emotional disturbances, or social retardation. In the dysarthric group will be found neuromuscular involvement of lips, mandible, tongue and palate. Accompanying this is usually found a history of eating difficulties or delay in good eating habits.

The speech of the spastic child is slow and shows poverty of movement, and often certain movements are made consistently so that their speech is more easily understood than that of the athetoid. The athetoid shows a variety of movements but few of these will be under his control. Clumsy, thick, and monotonous speech is often found in children with ataxia. A variety of tongue disturbances will be found, one of which is tongue thrusting—a distressing feature, because it can interfere very severely with feeding. Facial grimacing, if present, may involve the whole face or be confined to the mouth area only. Many of these children will show some deviation from the normal breathing pattern, and it is important that this type of difficulty should be given immediate attention. Protracted therapy is usually required, particularly in cases of reversed breathing.

Correction of poor breathing patterns will indirectly help voice disorders.

Hearing

The majority of the children undergoing hearing tests at the Cheyne Centre are under the age of seven. These tests are done as a routine measure, so that the appropriate training may be embarked upon as soon as possible. The speech therapist can prepare the child for the series of tests by practising conditioning. There are many difficulties here because a reliable response to a sound stimulus must be found. A child who is severely involved in all four limbs may have much difficulty in responding reliably. Therefore it entails careful observations to find a particular movement which he can make consistently and with as much ease as possible. These preparations are carried out by the teacher of the deaf and the speech therapist. When auditory training begins, the problems of head control and visual attention are again evident. In order to produce the optimum conditions a discussion between the teacher of the deaf, the physiotherapist and the speech therapist is extremely helpful.

In a very young deaf child, auditory training should be given primary consideration, because the acquisition of language, or at least understanding of language, which includes training in listening, is of vital importance.

Treatment

There are various methods of treatment available, each of them has something valuable to offer. These children vary tremendously in their difficulties, and it is therefore helpful to know and understand the principles of the various methods so that each child receives what is best suited to his needs.

Relaxation and correct positioning are essential during the building up of speech patterns. The speech therapist therefore looks for guidance from the physiotherapist in these matters and sometimes works with her. Speech is built up through a series of rhythmical sound patterns. It is the tune and rhythm of speech which is important to these children.

Many cerebral palsied children when first seen present a passive attitude towards people and their surroundings, and the desire to communicate is absent. The speech therapist can make good use of this 'silent period'. She has to create an enjoyable atmosphere for the child which gives him the opportunity of watching and listening to speech, and above all to enjoy speech activities. Thus the urge to contact his world through vocalisation or speech is being encouraged and aroused.

In all types of cerebral palsy the movements necessary for chewing, sucking and swallowing may be absent or limited, resulting in messy eating and poor table habits. These problems cause anxiety in the home, for a great deal of time may be spent over meals, and emotional attitudes develop which hinder the progress of good habits.

The occupational therapist and the speech therapist, working together, make an excellent team when tackling these problems. They observe the children during meal-times and give assistance where necessary. If possible, actual teaching and practice of new feeding habits should be done during a special period.

It is not always possible to help a child achieve a standard of speech which is socially acceptable. But even in these circumstances the speech therapist can find other ways to help the child communicate, and help him to enjoy the process of development in as normal an atmosphere as possible, and the gaining of confidence in his abilities no matter how limited they may be.

QUESTIONS

When asked what methods of relaxation she used, Mrs. Back said one needs to build up a technique for each child. The correct chair is very important where a sitting position is desirable. In some cases reflex inhibiting postures are useful.

BOOKS — NEW AND NOT SO NEW

THE ANATOMY OF JUDGMENT

By MRS. JOHNSON ABERCROMBIE

Reviewed by Dr. J. R. ELLIS, *M.B.E.*, F.R.C.P.

Physician to the London Hospital and honorary secretary to the Association for the Study of Medical Education.

The Anatomy of Judgment
By M. L. JOHNSON ABERCROMBIE, B.SC., PH.D.
London: Hutchinson, 1960, 149 pp., 25*s*.

Most teachers hope that what they say and show will be understood by the majority of their pupils. The degree of understanding may not be frequently assessed and not uncommonly testing of it is left to widely-spaced examinations. In these many other variables, such as memory, are involved, and failure on the part of the pupil to give the right answer is often attributed to his having forgotten it, while giving the wrong answer is frequently attributed to his lack of intelligence. The fact that he may have remembered accurately what he understood the teacher to mean is not often taken into consideration. Yet all teachers have had cause to discover, at one time or another, that their most precise and explicit words have had quite different meanings for different pupils.

Dr. Abercrombie, in the first part of this clearly written and very readable book, gives one reason why the same word, phrase or sight can mean different things to different people, or even to the same person in different circumstances. 'In receiving information from a given stimulus pattern we select from the total information available (that is, from the complex of the stimulus pattern in its context) and (also) from our own store of information.' The receipt of information therefore involves making a judgment, but in many cases—for instance, in seeing familiar things—this is done so rapidly and automatically that we are unaware of the extent of our personal involvement in the act, tending to regard the information as given.

Dr. Abercrombie's lucid exposition of the evidence for this statement is all the more interesting because it confirms what personal experience leads everyone to suspect. The main purpose of her book, however, is to consider the relevance of the statement to education, in particular to the interdependence of the processes of perception and reasoning. If 'many factors of which we are unconscious influence our judgments, both in cases where we are not aware of making any (as in seeing) and in those where we are (as in evaluating evidence from an experiment)', then we might 'make more valid judgments if we could become conscious of some of these factors.'

Dr. Abercrombie describes a teaching course which she gave to preclinical medical students of University College, London. The course consisted of eight free group discussions, concerned with seeing, language, classification, evaluation of evidence, causation and a review of the course itself. On each occasion each student first re-

corded his own interpretation of a particular statement or visual stimulus: and then, in discussion, became aware not only that his fellows had made different interpretations, but also to some extent of the reasons for his and their choice of interpretation. 'The main difference between this and traditional methods of teaching is the amount of attention that is paid to the *processes* of observing or thinking, as distinct from the results.' 'What the student learns, it is hoped, is not only how to make a more correct response when he is confronted with a similar problem, but more generally to gain firmer control of his behaviour by understanding better his own ways of working.'

Dr. Abercrombie set out to evaluate the success of her project by comparing (over three years) the performance in observation tests of students who had taken the course with that of students, otherwise similar, who had not yet taken it. She found that students who had taken the course did significantly better than the others in four respects—'they tended to discriminate better between facts and conclusions, to draw fewer false conclusions, to consider more than one solution to a problem, and to be less adversely influenced in their approach to a problem by their experience of a preceding one.'

Here then is something which is paradoxically rare in an age when more attention is paid to and more hope based on education than ever before—an investigation into learning processes *and* a successful experiment in teaching. That a critical faculty can be acquired is confirmed, and a method whereby it can be cultivated is shown. The method is time-consuming and somewhat uncomfortable for both teacher and students. For the purpose of the experiment Dr. Abercrombie learnt a few simple skills that can be applied to teaching. 'The most important of these,' she writes, 'is listening, and the second is tolerating the expression of hostility in a relationship in which it is customarily suppressed.' These words alone show why her book should be read by all those who, while believing that education is important, fondly imagine that teaching is synonymous with talking. One might add that the book also demonstrates the value of a simple rule that can be, but is not always, applied to teaching—that the teacher knows precisely what he is aiming to achieve before setting out to achieve it.

Certainly it should be read by all those engaged in medical education, for their primary purpose is to inculcate both the power of accurate observation and a critical faculty in their students. They should counter any suggestion that the method of teaching described is expensive of time and money, by asking themselves what alternative methods are in use (at what cost) and by attempting to assess the importance of making doctors 'more objective and more flexible in their behaviour'. Dr. Abercrombie points out that a statement by a committee of the Royal College of Physicians served as a text for her guidance in this work. 'The average medical graduate "tends to lack curiosity and initiative; his powers of observation are relatively undeveloped; his ability to arrange and interpret facts is poor; he lacks precision in the use of words".' The statement was made in 1944—eleven years later another committee of the same College published a further report in which it clearly saw no reason to deviate from this statement. Whether or not any improvement can be reported in 1964 must depend largely on there being further studies of the kind described in this book, and on the degree to which their results are applied to teaching practice.

On such work the future of all education depends, for otherwise it can never rise much above what it unhappily is so often today, no more than the blind acquisition of knowledge, without the ability to use it or to discriminate between true and false. It is a frightening thought that education has, by conferring on many people the gift of reading and writing, increased the quantity of ignorance which is written and read. Mercifully it cannot all be remembered.

JOHN R. ELLIS

Recent Neurological Research

Edited by A. BIEMOND.

Amsterdam: Elsevier, 1959, 330 pp., 47*s.* 6*d.*

The title of this book is a little misleading. It is in fact published to celebrate the 50th anniversary of the Amsterdam Neurological Society and consists of 28 short papers by Dutch neurologists. These cover a wide field, ranging from cerebellar degeneration and echoencephalography to the theory that the interfibral vacuoles of the cerebral cortex are a special organ promoting memory, and the hodology of the elephant's spinal cord. Each article is hardly detailed enough to be of great interest to others than pure neurologists, but for those of us who know and admire the neurologists of the Netherlands it is of considerable interest to see the subjects in which they are specially interested.

N. S. ALCOCK

Modern Ideas of Physical Education

By M. W. RANDALL.

London: G. Bell & Sons, revised edition, 1959, 168 pp., 10*s.* 6*d.*

Here is perhaps the most up-to-date account of physical education available today. The title '*Modern Ideas*' should not, however, be taken to imply that the author is particularly forward-looking. Rather he is describing what actually goes on at present: and in addition he gives a most adequate and commendably catholic bibliography for further reading.

In his description of what is now being done he has avoided mentioning modern dancing, which plays an increasing part in the girl's physical education; and indeed, as is the case with many male physical education teachers, he frankly states that he is not in sympathy with the idea of modern dancing for boys. Certainly, if physical education is to have a sound scientific basis, far more clarity of description both of aim and of method must be demanded from its exponents.

In his general educational comments, Mr. Randall has little to say which will not be gained from the first year of any teachers' training course, but his book has rightly had a wide sale in its first edition, as providing a good coverage of the subject for the general reader. Mr. Randall has compressed a vast field into a small space without at any point losing in readability.

A large proportion of the book is devoted to a consideration of *Posture*, and here he is not so helpful. His physiology is not up to date, and even though the coverage is general, it should be accurate in a book of this sort. He grudgingly admits that there may be something in the pioneer work of Mathias Alexander; but Alexander's work has completely revolutionised the approach to postural re-education, and it will inevitably play an increasing part in physical education, long after the current enthusiasm for 'the Art of Movement', circuit-training, and muscular hypertension has had its day.

In spite of this criticism, Mr. Randall's book can be recommended to anyone who requires a short and intelligent account of what is being done in physical education.

WILFRED BARLOW

The Principles of Exercise Therapy

By M. DENA GARDINER, F.C.S.P.

London: G. Bell & Sons, *2nd ed.*, 1959, 289 pp., 21*s.*

In assessing and treating any abnormal physical condition, one becomes increasingly aware of the need for more analytical observations on normal activity. Miss Gardiner's book provides many facts of interest to both students and veterans in this field.

The new edition contains a chapter by Miss Martin Jones on *Proprioceptive Facilitation*, which is the basis of a treatment technique developed by Dr. Kabat and Miss Knott at the Kabat-Kaiser Institute in California. In introducing this technique, Miss Martin Jones reminds us of Hughlings Jackson's statement that: 'Motor centres know nothing of muscles: they only know

of movement'. The method replaces original isolated muscle group exercises by patterns of movement based on natural ones, and these are facilitated by stimulation of the proprioceptors. Unfortunately, the major part of this chapter deals with detail of starting position and grasp, and the theory is not discussed. However, it makes us wonder whether we have not hitherto oversimplified muscle re-education. In the field of cerebral palsy, where these pathways are not much used, it is certain that too little has been tried on these lines.

Miss Gardiner dismisses some ideas and findings too readily. Although the book is primarily written for students, one would like to have seen greater elaboration on weight-lifting, for instance, and a more detailed assessment of the fatigue limit in relation to what Miss Gardiner maintains are the five essential factors in muscular efficiency—power, endurance, volume, speed of contraction, and co-ordination. Small wonder that the problem of cerebral palsy remains so complicated!

The book is certainly valuable to all concerned in the field of muscle activity, and one hopes that it is merely the *hors d'oeuvre*, to be followed by a further course of food for thought.

PATRICIA BEAMAN

Psychosomatics

Edited by J. BOOIJ.

Amsterdam: Elsevier, 1957, 125 pp., 24s.

Psychosomatics represents an old approach to patients which has lately been remarkably rejuvenated and invigorated by new methods. These methods are bringing it into line with other aspects of modern scientific medicine.

This small book contains five lectures given in 1954 by invited speakers to a joint meeting of the Dutch Psychiatric and Neurological Society with the Association of Dutch Internists. The lecturers include some (J. Groen and J. Bastiaans, for example) whose names are well known outside their own country for their impressive contributions to psychosomatic theory and practice. The chapters outline some of the possibilities and limitations of psychosomatic medicine; aspects of syndrome shift and suppression; the relationship with anthropology; the psychopathology of peptic ulcer; and some results of psychotherapy.

The lecturers are scrupulously fair in not attempting to force all illness into the psychosomatic ambit. Rather a long time has elapsed since the lectures were delivered, but they give a useful sketch of a rapidly developing subject for those coming new to it.

JOHN APLEY

Lectures on Epilepsy

Edited by LORENTZ DE HAAS.

Amsterdam: Elsevier. *London:* van Nostrand, 1959, 172 pp., 24*s*.

The 75th anniversary of the Epilepsy Centre, 'Meer en Bosch', at Heemstede in the Netherlands, was celebrated in 1957. Various eminent speakers were invited to mark the occasion, and four of their lectures are now published under this title. The psychiatric aspects of epilepsy have received most attention in a volume in which the views expressed by each author are very personal, and which is not intended to be a review of its subject or a textbook.

This is clear from the first paper, '*Considerations on Temporal Lobe Epilepsy and its Surgical Treatment*', by Professor David and Dr. Dell (Paris). They consider that a temporal lobe seizure is a form of generalised epilepsy rather than an attack of focal origin, and the anatomical lesions observed in cases of temporal lobe epilepsy are regarded as non-specific. However, no mention is made of those patients in whom neoplasms or hamartomas have been demonstrated on postoperative examination of the excised lobe, in some of which the macroscopic appearances are almost normal, and it is interesting that no mention is made of the value of pneumoencephalography in the study of temporal lobe

seizures. The fact that electroencephalographic abnormalities are often fairly localised is attributed to the lower convulsive threshold of rhinencephalic structures. The authors consider electrocorticography and cortical stimulation of little value, and they regard the use of indwelling electrodes as morally unsupportable. However, they advocate the use of subcortical leads introduced by a sterotaxic apparatus. This lecture would be more valuable if many more references to the literature were given in support of the arguments. But it is useful to read a paper which is at variance with the more generally accepted principles, in order that these may be reassessed and criticised, whether or not the alternative offered proves acceptable.

The second lecture, on '*Psychic Disturbances in Epileptics*' is by Dr. Vislie and Dr. Henriksen of Oslo. Their series comprises 162 patients, but the absence of data about the clinical condition makes it difficult to evaluate their material. Insufficient attention is paid to differences of pathology, the analysis being mainly symptomatic and, to a limited extent, topographical. For instance, 25 of 142 patients who had pneumoencephalograms showed diffuse atrophy, but we are not told their diagnosis. Later we read 'in the group with known aetiology (of epilepsy) there was an excess of males, mainly due to the higher incidence of severe head injuries among males.' Clearly, the scattering of severe head injuries among any group of epileptics can considerably influence the conclusions. There is an excellent bibliography which would be of value to others studying the subject.

The third lecture, by Dr. Landolt of Zürich, gives his views on 'forced normalisation' in epileptic patients who show psychotic episodes and schizophrenic attacks. These views are of considerable interest, and, though perhaps not generally accepted, they are stimulating and will cause others to consider them in the light of their own experience. More EEG tracings would have been useful.

The final paper comes from the Epilepsy Centre itself and is by the editor and Dr. Magnus. Their subject is '*Episodic Mental Disorders*', and in this lecture an attempt is made to reclassify some twilight and dysphoric states and psychotic conditions in the light of EEG correlates. This is bound to be controversial, and some points are at variance with the first lecture.

These papers provide a concise English summary of the views of each author, and the book should therefore prove a useful work of reference.

PETER H. SCHURR

NOTICES

ACTION FOR MENTAL HEALTH

ANNUAL MEETING OF WORLD FEDERATION FOR MENTAL HEALTH

Edinburgh, August 8–13, 1960

THE Federation is holding its 13th Annual Meeting in Edinburgh this year, by invitation of the Scottish Association for Mental Health. It will start in the afternoon of Monday, the 8th, and end on the following Saturday afternoon, the 13th. The main meeting-place will be the McEwan Hall, and adjacent University buildings.

The programme will be based on the six principal points around which the activities of the World Mental Health Year are focused: surveys in the field of mental health and ill-health; mental health in education, including professional training; industry and mental health; mental health and migration; and ageing. There will be special sessions on mental deficiency, and it is hoped that the meeting will be attended by people participating in the Conference on the Scientific Study of Mental Deficiency. The Executive Board select the speakers very largely from among those who have signified their intention of attending the meeting, including representatives of as many professions and cultures as possible. They will all be experts in their specialty. The meeting will be conducted in English and French, simultaneous interpretation in these languages being supplied for the main sessions.

Further particulars from: Miss Esther M. Thornton, Secretary-General, W.F.M.H., 19 Manchester Street, London, W.1.

MANAGEMENT OF CEREBRAL PALSY

B.C.W.S. AND DURHAM UNIVERSITY CONFERENCE

Newcastle upon Tyne, July 8–9, 1960

THIS conference has been arranged by the British Council for the Welfare of Spastics, in co-operation with the University of Durham, and it will include sessions on the work being done at the Percy Hedley School and Clinic for Spastics in Forest Hall, which is 4½ miles from the centre of Newcastle. The conference will meet at the Ethel Williams Hall, a hall of residence of the University which is a few minutes' walk from the School. Admission is by invitation only, and the attendance is being limited to 40, to allow everyone to speak in the discussions. On the first day, Prof. R. S. Illingworth will speak on the early diagnosis of cerebral palsy and show a film; Dr. Mary D. Sheridan of the Ministry of Health will read a paper on the ineducable child with cerebral palsy; Dr. E. Ellis, medical director of the Percy Hedley School, will talk about cerebral palsy 1953-1960; and Miss D. M. Peaps, the School's almoner, will discuss school-leavers, adolescents and adults with cerebral palsy. There will be a demonstration by the staff of the School in the evening. Next day Dr. E. M. Creak will read a paper on the emotional implications of cerebral palsy; Dr John N. Walton will read one on clumsy children; and Mr. J. Hankinson, F.R.C.S., one on the place of neurosurgery in cerebral palsy. The Chair throughout the conference will be taken by Prof. S. D. M. Court, Sir James Spence professor of child health in the University.

Particulars from the secretary, B.C.W.S., 13 Suffolk Street, S.W.1.

MATURATION

FIRST EUROPEAN CONGRESS OF CHILD PSYCHIATRY

Paris, September 16–20, 1960

PROFESSOR MICHAUX will preside over this Congress of Child Psychiatry, to be held at the New School of Medicine, 45 rue des Saints Pères, Paris VIe. The discussions will cover the biological basis, stages, and social implications of maturation, and the Congress will end with a symposium on child psychoanalysis.

Particulars from the Secretary-General, Dr. D. J. Duché, Pavillon Clérambault, Hôpital de la Salpétrière, 47 Boulevard de l' Hôpital, Paris XIIIe.

FIRST INTERNATIONAL CONFERENCE ON CONGENITAL MALFORMATIONS

London, July 18–22, 1960

THE National Foundation of the U.S.A. is sponsoring this conference at Church House, Westminster, under the general chairmanship of Prof. J. D. Boyd, with Basil O'Connor as president, Dr. Morris Fishbein and Melvin A. Glasser as vice-presidents, and Sir Geoffrey Marshall as an honorary president. The 24 members of the International Medical Congress Ltd., who have announced the conference, include Prof. Robert Debré from Paris, Prof. Gino Frontali from Rome, Dr. Renato Bomfim from Sao Paulo, Brazil, Prof. Arvid Wallgren of Stockholm, Dr. Carlos Ottolenghi of Buenos Aires, and Dr. Frank L. Horsfall junior, Prof. Charles Ragan, Prof. Edward L. Tatum, Dr. Norman H. Topping, Dr. Josef Warkany and other distinguished Americans. The main subjects for discussion will be: (1) incidence; (2) intrinsic factors (genetics); (3) extrinsic factors (environment); (4) general developmental mechanisms; (5) abnormal developmental mechanisms; (6) maternal-foetal interactions; (7) physiological and medical problems; and (8) perspectives. There will be simultaneous interpretation of the addresses into English, French and German, as required.

Particulars from The Secretariat, at 67 New Bond Street, London, W.1.

LONDON CONFERENCE ON THE SCIENTIFIC STUDY OF MENTAL DEFICIENCY

London: July 24–28, 1960

THIS Conference, which will be held at the British Medical Association's headquarters in Tavistock Square, London, W.C.1, is intended for professional workers in all branches of the mental deficiency field—medical, psychological, educational, social and administrative. It is sponsored by the American Association on Mental Deficiency, the Royal Medico-Psychological Association, the Royal Society of Medicine, and the British Psychological Society, in co-operation with the National Association for Mental Health in London. It is being held as a contribution to the World Mental Health Year, and the subjects for discussion will include biology and genetics, pathology and biochemistry, psychopathy and behaviour problems, physical therapy, learning problems, epilepsy and cerebral palsy, and many kindred subjects.

Further details can be obtained from the joint hon. secs.: Dr. A. Shapiro and Dr. A. D. B. Clarke, at the conference office, 39 Queen Anne Street, London, W.1.

SOCIETY FOR MUSIC THERAPY AND REMEDIAL MUSIC

MANY attempts are now being made to use music as a therapeutic agent in the treatment of illness of mind or body. The development of 'music therapy' will first depend on the study of the nature and scope of this new subject, in order to formulate sound general principles and methods of application.

The Society for Music Therapy and Remedial Music has recently been formed to promote the use of music in the treatment, education, training and rehabilitation of children and adults suffering from emotional, physical or mental handicaps. Its members include doctors, musicians, psychologists, educationists, teachers, social workers, therapists, nurses and students; as well as official and academic bodies such as the Ministry of Education, the National Association for Mentally Handicapped Children and the University of Durham.

The Society's activities are extensive—its members hold frequent meetings with lectures, talks and group discussions; it is an active information centre, publishes a *Bulletin* several times a year containing summaries of the lectures, news and useful information; it issues pamphlets on various aspects of music therapy; collects unpublished papers containing data and observations; and it gives advice to students on their work and provides facilities for them to visit some hospitals or special schools.

The Society is taking an active part in World Mental Health Year 1960, with a programme of lectures on '*Music and Mental Health*' in all parts of the country, and it is to hold a conference on April 30 in London on '*Music Therapy and the Education of the Child*', with emphasis on mental health. The conference will discuss the cerebral palsied child, among others.

The rapid and steady growth of this young society owes much to the active support it is receiving in medical, musical and educational circles, and to the interest music therapy is evoking among people concerned with the health and welfare of handicapped children and adults.

Juliette Alvin

NOTE: This is by no means the only music therapy society in this country. Others are the Council for Music in Hospitals (5/6 Raymond Buildings, Gray's Inn, W.C.1), which organises concerts at mental and other hospitals, and the Association for Music Therapy in Hospitals (50 Belgrave Road, London, S.W.1), which encourages music therapy in mental and other hospitals and runs a training scheme for music therapists. Details of these will appear in future issues.—EDITOR, *Cerebral Palsy Bulletin.*

ABSTRACTS

IN COLLABORATION WITH "*Abstracts of World Medicine*," PUBLISHED BY THE BRITISH MEDICAL ASSOCIATION, AND WITH THE KIND COLLABORATION OF THE EXCERPTA MEDICA FOUNDATION AND "*Obstetrical and Gynecological Survey*".

Structural Alterations in the Cerebellum in Cases of Cerebral Palsy. Their Relation to Residual Symptomatology in the Ataxic-Atonic Group

C. B. COURVILLE. *Bulletin of the Los Angeles Neurological Society*, Sept. 1959, **24,** 184–165.

At the Cajal Laboratory of Neuropathology, Los Angeles County Hospital, the author has investigated post mortem the incidence of cerebellar lesions (and the relationship of such lesions to symptoms) in cases of cerebral palsy. Of 126 such patients who had survived for at least one year cerebellar lesions were found in 8, of which 5 are described in some detail. The typical picture was of a cerebellum which appeared normal macroscopically, but on section showed diffuse cortical atrophy, with selective loss of Purkinje and granule cells, later followed by degeneration of corticofugal nerve fibres and by gliosis. In cases in which gross cerebellar lesions such as generalised atrophy and diffuse cortical sclerosis were present the cerebrum was similarly affected. In no case was cerebellar damage alone found, all showing also major lesions in the cerebrum. Only one patient, a child aged 16 months, presented with pure cerebellar symptoms, manifested by generalised atonia. In all the other patients any cerebellar symptoms were overshadowed by the motor and mental manifestations of the cerebral lesions.

The author points out that the widespread and uniform nature of the cerebellar lesions indicates an anoxic origin, although embryological, traumatic, and inflammatory causes are other aetiological possibilities. He suggests that closer study be made of cerebellar lesions in cases of cerebral palsy, and if ataxic-diplegic cases were differentiated from atonic-ataxic cases at clinics, the disappointing response to treatment in the latter would be better understood; by the nature of their predominating cerebellar lesions these patients are quite resistant to the usual measures of physical therapy directed towards their rehabilitation. *Janet Q. Ballantine*

Activity of Glutamic-oxalacetic Transaminase and Lactic Dehydrogenase in Cerebrospinal Fluid and Plasma of Normal and Abnormal Newborn Infants

M. LENDING, L. B. SLOBODY, M. L. STONE, R. E. HOSBACH, and J. MESTERN. *Pediatrics*, Sept. 1959, **24,** 378–388.

This paper from New York Medical College and the Flower and Fifth Avenue Hospitals, New York, reports a study of the activity of the enzymes glutamic oxalacetic transaminase (GOT) and lactic dehydrogenase (LDH) in the serum and (CSF) of 54 healthy newborn infants, aged 2½ hours to 10 days, and 20 newborn infants, aged 2 to 115 hours, with suspected intracranial birth injury. In the normal infants the plasma GOT activity ranged from 5 to 102 units per ml. per minute (average 36.9 ± 18.9) and the plasma LDH ranged from 440 to 2,540 units per ml. per minute (average $1,165 \pm 543$); the values for the CSF were: GOT 1 to 10 units per ml. per minute (average 4.5 ± 2.2) and LDH 3.3 to 120 units per ml. per minute (average 32.3 ± 24). In the infants with intracranial injury the GOT activity of the CSF showed an increase of 82% over that of normal infants and the plasma GOT an

increase of 18%. The LDH activity of the CSF showed a mean increase of 309% over the normal but the plasma LDH showed an increase of only 11% over normal. It is suggested that the activity of these enzymes in the cerebrospinal fluid may be helpful in the study of infants in whom intracranial birth injury is suspected.

Winston Turner

Effect of 'Ciba 13.155' in the Treatment of Spastic Paraplegia. (In English.)
E. PEDERSEN and P. SCHLEISNER. *Acta psychiatrica et neurologica Scandinavica*, 1959, **34**, 342–353.

Ciba 13.155 is a polysynaptic inhibitor of the central nervous system, its chemical formula being 2-hydrazino-4:6-bis-diethyl-amino-1:3:5-triazine dihydrochloride. At the Kommunehospital, Aarhus, Denmark, the drug was used in 50 patients with various spastic disorders such as disseminated sclerosis, hemiplegia, and subacute combined degeneration of the spinal cord. At first it was given in doses of 50 mg. 3 times a day by mouth. Subsequently the optimum dosage was found to be in the region of 6 mg. per kg. body-weight per day. The gait improved in 29 of the cases, but the most surprising and unexpected result was the improvement in bladder control of 12 patients with frequency and incontinence. The most frequent side-effects were mild heartburn and epigastric pain, which could probably be avoided by coating the tablets of the strongly acid drug. It would seem that this substance may prove to be the most useful so far produced for the relaxation of muscular tone in spastic disorders.

G. S. Crockett

Flexion-spasm Disease
G. B. CAVAZZUTI. *Lattante*, Aug. 1959, **30**, 441–456.

This paper starts with a brief summary of recent work on flexion spasm and then describes the results obtained in 15 children treated with anticonvulsant drugs such as 'mysoline', (primidone), 'dintoina', or 'luminal' (phenobarbitone) together with ACTH (corticotrophin) at the Paediatric Clinic of the University of Modena. Eleven children were found to derive little permanent benefit. Some were unaffected by the treatment, while others showed varying diminution in the length and severity of the attacks. However, the remaining 4 children appeared to be cured, 3 of these having been treated with anticonvulsants and the fourth with prednisone. The delay in psychomotor development remains long after the spasms have ceased; thus in one patient it was 2 years before a normal level of development was reached. No common origin for the attacks could be found. There was some indication that the prognosis was best in those patients who responded early and in a well-marked way to therapy.

J. G. Jamieson

Flexion Spasms and their Treatment with ACTH and Hydrocortisone.
G. DUMERUTH. *Helvitica paediatrica acta*, Sept. 1959, **14**, 250–270.

From the University Paediatric Clinic, Zürich, the author reports that as the result of treating 10 children, all but one under 3 years of age, suffering from typical attacks of flexion spasm with ACTH or hydrocortisone 6 were definitely benefited by the treatment, but the other 4 showed no improvement, nor did 2 atypical cases of the disorder. Neither the history nor the clinical and laboratory findings pointed to any obvious cause for the difference in results.

In the children who improved the EEG became more nearly normal, they became quieter, showed more co-ordination in their movements, and in one case particularly the retardation in development became less marked. A follow-up report states that after 3 months' intermittent therapy one child had become free of attacks, though a skull injury had caused a temporary relapse, while a second case after 4 months' therapy showed both clinical improvement and the absence of signs of epileptic activity in the EEG. Side-effects were not serious; most of the children developed a plethoric appearance of the face and a much larger

appetite, and 2 developed a temperature which could not be attributed to infection.

When ACTH was given as a first treatment all other anticonvulsant drugs were withheld, but when improvement set in an anticonvulsant also was prescribed. Intermittent therapy is considered essential, since relapses do occur, and the long-continued administration of large doses of cortisone may provoke grand mal seizures.

J. G. Jamieson

Neuro-psychological Disorders in Cases of Congenital Cardiac Disease. (In Russian.) V. V. KOVALEV. *Z. Nevropat. Psihiat*, 1959, **59**, 986–993.

The author reports the principal neurological and psychiatric findings in 86 cases of congenital cardiac disease and attempts to relate the majority of the disorders to oxygen lack. Of these patients, 54 were aged between 3 and 16, and 32 between 16 and 36 years. The main cardiac defects were Fallot's tetralogy (29 cases), patent ductus arteriosus (15), septal defects (9), and Eisenmenger's complex (5), while 42 cases were of the cyanosed type and 44 were non-cyanosed.

Slight neurological abnormalities consisting in mild cranial nerve palsies, poor motor co-ordination, and weak sensory perception were present in 64 cases, and evidence of neurovascular disturbance was found in 28 cases, these patients having severe spasmodic headaches, with pallor and vomiting. In regard to psychological abnormalities the predominant finding (82 cases) was of some degree of asthenia, these cases falling into three groups:

(1) The 37 with purely neurasthenic features of gradual onset. These patients complained of fatigue on the slightest exertion, had poor mental concentration, and disturbed sleep, nightmares with a 'suffocation' theme being common. Moods were variable, being either euphoric or apathetic or depressed, extremes of euphoria or apathy being noted in those in whom oxygen lack was most marked. Some patients tended to be egocentric and showed lack of normal emotion. Children were older in appearance than their years and seldom displayed enthusiasm or affection. Hypochondria and apprehension about the course of the illness were not features of this group.

(2) The 36 patients who showed neurasthenia with a variable degree of mental retardation, which in many had been noted from a very early age; there was one case of imbecility. In 7 cases, however, mental retardation did not become apparent until the age of 10 to 12 years.

(3) The notable abnormality in the third group (9 cases) was the psychotic features. In many of these patients irritability, restlessness, and aggressiveness had been noted in early childhood; threats of violence were, however, rarely translated into action. Grandiose ideas were a feature of patients in this group.

In all the cases studied the blood oxygen content and pulmonary gaseous exchange were determined. Reference is made to the literature dealing with the effects of experimental anoxia on cerebral activity, and it is pointed out that the reported effects were very similar to many of the neurasthenic features found in these patients with congenital cardiac disease. In those showing mental retardation this was usually associated with physical abnormality and is considered to have been due to congenital cerebral abnormalities. The character defects seen in Group 3 were no doubt enhanced by oxygen deficiency, but may have been primarily due to faulty upbringing.

Margot G. Dunlop

Tests for Phenylketonuria: Results of a One-year Programme for Its Detection in Infancy and among Mental Defectives
N. K. GIBBS and L. I. WOOLF. *British Medical Journal*, Sept. 26, 1959, **ii**, 532–535.

Early detection and treatment of phenylketonuria are essential if irreversible mental deterioration is to be avoided. A scheme which aimed at the early detection of cases of phenylketonuria was tried in Cardiff, mothers of infants born in the city in the year March 1, 1958, to March 1, 1959, being asked for a fresh specimen of the

infant's urine when the child was 3 weeks old.

Although there were 4,530 live births in Cardiff during the year, only 1,192 urine specimens were collected. All were tested with ferric chloride and many of them also with 'phenistix', a test strip which, when dipped in urine, turns green in the presence of phenylpyruvic acid. Of the 1,192 specimens, 51 were strongly alkaline and were rejected; of the remaining 1,141 specimens, one gave a positive reaction for phenylpyruvic acid.

The problems of such mass schemes for the early detection of phenylketonuria are discussed. It is suggested that the use of phenistix provides a simpler, quicker, and more specific method of testing urine than any hitherto employed. *H. Harris*

The Development of Ten Children with Blindness as a Result of Retrolental Fibroplasia: a Four-year Longitudinal Study

A. H. PARMELEE JR., C. E. FISKE, and R. H. WRIGHT. *A.M.A. Journal of Diseases of Children*, Aug. 1959, **98**, 198–220.

The 10 prematurely born children, blind as a result of retrolental fibroplasia, with whom this paper from the University of California Medical Center, Los Angeles, is concerned, were the subject of a previous investigation (Parmelee, *A.M.A.J. Dis. Child.*, 1955, **90**, 135), in which their development during the first year of life was compared with that of 10 prematurely born infants with normal vision and 80 full-term infants. The Gesell infant test was used, and the mean scores for the three groups showed no statistical difference, indicating that at that stage the blind children were developmentally normal.

A review of their progress 4 years later is now presented. The tests used were the Vineland Social Maturity Scale and the Stanford-Binet Form L. The I.Q. was over 80 in 6 cases and below 80 in 3, while one of the children who was found to be deaf as well as blind and who could not therefore perform the tests was assessed as being of normal mental potential from the observations of 5 independent observers. The 3 children functioning at a mentally retarded level were considered to have normal mental potential, but performance was suppressed by emotional problems. Electroencephalograms were recorded in 6 cases, in 2 of which they were abnormal in that they showed more waves at 1 to 3 c.p.s. than occur in normal children of the same age group. One of these abnormal tracings was from one of the 3 retarded children, those of the other 2 being normal.

Although all 10 children were originally certified as blind, 3 have since developed useful although very limited vision in one eye, and another has developed sufficient vision to read large print with glasses. The 3 retarded children were amongst those with no vision at all. The main body of the paper is devoted to fairly detailed biographies of the 10 children. The authors also discuss (with considerable insight) the problems of the blind child and his parents.

(Every paediatrician with continued responsibility for the care of the premature child with retrolental fibroplasia will be interested in this richly descriptive paper.) *David Morris*

Post-encephalitic Behaviour Disorder—a Forgotten Entity: a Report of 100 Cases

S. LEVY. *American Journal of Psychiatry*, June 1959, **115**, 1062–1067.

The author emphasises the importance of an accurate differential diagnosis between ordinary juvenile 'behaviour disorders' and those due to organic brain damage following encephalitis. He claims that the diagnosis can be easily made provided the possibility of a post-encephalitic syndrome is borne in mind, when the following outstanding signs are usually present: (1) hyperkinesis with either choreiform or tic-like movements; (2) inability to maintain quiet attitudes; (3) clumsiness of movement; (4) explosive motor release of all voluntarily inhibited activities. The aetiology usually includes a history of brain damage by high forceps delivery, or of a severe infectious disease in infancy, or frequent attacks of hyperthermia without

apparent cause. The parents' description of the disorder shows that it is expressed almost entirely in the volitional sphere. The child is overactive, restless, shows a short attention span, is unpredictable, destructive, and usually incapable of showing remorse. School reports confirm the child's 'inability to apply himself'. Physical and neurological examination are usually negative, but recently the photo-metasol threshold test has shown significant differences between hyperkinetic and non-hyperkinetic children. The I.Q. is usually normal or higher and psychological testing does not contribute to the aetiology. The child is intolerant of frustration and seems unable to delay gratification of his demands.

Some 20 years ago amphetamine sulphate was introduced for the treatment of this disorder and the results in the series of 100 such patients here presented again show the effectiveness of the drug. It was given in doses of 10 to 40 mg. daily either in a single dose in the morning or with a second 'booster' dose at lunch-time half the strength of the first. In some cases the beneficial effect was delayed for 2 or 3 weeks, but once apparent it was spectacular. Restlessness and irritability decreased and the child's concentration span increased markedly. The patients became less 'driven' in their behaviour and their parents and teachers responded much more favourably. Side-effects were few. In one case the child was either very sensitive to the drug or the diagnosis was at fault, as he became more disturbed over a long period, while in another case the patient developed a delayed dermatitis, but these were the only two in which the drug had to be discontinued. The author recalls that the drug must be given over a prolonged period, and recommends that parents should be warned that during the first 2 weeks of treatment the child's behaviour reactions may become exaggerated; however, provided that these reactions do not persist for more than 3 weeks this is a good prognostic sign. In view of the efficacy of amphetamine sulphate in the treatment of this disturbing and chronic illness it is particularly important that behaviour disorder when due to encephalitis should not be treated as a psychogenic illness.

M. R. Medhurst

The Respiratory Distress Syndrome and Its Significance in Premature Infants

W. A. BAUMAN. *Pediatrics*, Aug. 1959, **24**, 194–204.

The significance of the respiratory distress syndrome in premature infants was studied in 201 patients admitted consecutively to the Premature Nursery of the Babies Hospital, New York, between December 1956 and March 1958. Each infant was examined every 6 hours for the first 72 hours of life for signs of respiratory distress, which were assessed according to the criteria of Silverman and Andersen, a score being given for the following physical signs: (1) see-saw movement of the chest, the sternum moving inwards as the abdomen moves out; (2) inspiratory indrawing of the intercostal spaces; (3) inspiratory retraction of the ziphoid cartilage; (4) descent of the chin on inspiration; and (5) an expiratory grunt. Each sign was scored 1 if present and 2 if marked; the maximum 'retraction score' was therefore 10, and a score of 2 or more was considered to indicate the presence of the respiratory distress syndrome.

The incidence of significant signs varied inversely with the birth weight and diminished throughout the first 24 hours of life. Of the 201 infants 37% had significant retraction in the first 6 hours of life and 19% at 24 hours. Of 43 infants who died in the first week of life 33 had scores of 2 or more and 20 of these had hyaline membrane in the lungs; of the 10 infants who had no signs of respiratory distress only one had a hyaline membrane. Approximately three-fifths of affected infants died in the first week of life, compared with about one-tenth of those not affected.

The author concludes that the retraction score is a valuable prognostic aid, but since 40% of infants with clinical signs of respiratory distress survive without

specific treatment any proposed therapeutic measures must be critically evaluated before they can be considered beneficial.

H. G. Farquhar

Prolonged Neonatal Anoxia without Apparent Adverse Sequelae

C. CHARLTON MABRY. *Journal of Pediatrics*, 1959, **55,** 211. (See *Obstetrical and Gynecological Survey*, 1960, **15,** 69.)

The capacity of foetal and neonatal subjects to withstand greater degrees of hypoxia and for longer periods than adults is well recognised. The longest recorded times that newborn infants have survived without apparent adverse sequelae are 14 minutes without breathing (*Brit. Med. J.*, 1955, **i,** 1071) and 30 minutes without a cord pulse (*Lancet*, 1958, **i,** 999). In each instance, however, the presence of a demonstrable heartbeat provided the encouragement to continue resuscitation. These infants cannot, therefore, be considered to have survived 'total anoxia'. The present report concerns a prematurely born newborn infant who survived a long period of asphyxia with no demonstrable heartbeat. Although the duration of 'total anoxia' was longer than has been recorded for any surviving infant, no adverse sequelae are apparent at this time.

A Negro boy, now 20 months of age, was born prematurely to a 19-year-old primigravida after a gestation period of approximately 35 weeks. The mother's labour was spontaneous and precipitous, being accomplished with the aid of 'low forceps'. No antepartum sedation or analgesia was used. Fetal heart sounds were heard until near the time of delivery and a viable infant was expected. However, an infant weighing 3 lb. 4 oz. was delivered who had no manifestations of life; that is, no heartbeat, respirations, pulsation of the umbilical cord, or movement of voluntary muscles. Resuscitation was started immediately, using a mouth-to-endotracheal-tube technique wherein the operator breathes oxygen-enriched air and in turn rhythmically inflates the infant's lungs via the endotracheal tube. This procedure was carried out for 15 minutes, at the end of which time he was declared 'stillborn' because no sign of life had been or could be detected.

The infant was prepared for the morgue and was wrapped in a shroud. However, instead of being delivered to the morgue, the body was taken to a nearby room for demonstration of resuscitative techniques. The demonstration was started 25 minutes after the infant was born. An endotracheal tube was inserted and the lungs were inflated by the expired air of three separate operators. Thirty minutes after delivery a pulse was first detected in the umbilical cord. A stethoscope was then placed on the baby's chest and a slow, irregular, weak heartbeat was heard for the first time. Resuscitation was continued with the technique initially used and 30 mg. of caffeine sodium benzoate was administered intramuscularly. The infant first gasped at the age of 40 minutes and began to breathe regularly and spontaneously at the age of 60 to 65 minutes.

The baby was then transferred to an Isolette with oxygen (38 volumes per cent) and received additional supportive care consisting of gavage feeding and antimicrobial therapy (chloramphenicol). After the first week, special premature infant care became unnecessary because the infant was 'hardy', and his course during the remaining five weeks in the nursery was characterised by steady growth and development. At no time did he have seizures or detectable jaundice.

The infant's growth has been consistent and adequate in relation to his birth weight; at 20 months of age his weight and length are within the 25th and 50th percentiles, respectively. He appears to be a normal toddler without any neurologic defects. He is a calm child with no abnormal motions of his hand and with no hyper- or hypotonia. He developed a social smile at 2 months of age, held his head steady at 3½ months, sat alone at 8–9 months, pulled to a standing position at 11–12 months, walked alone at 15 months, and used four to five words at 16–17 months of age.

[The Editor of *Obstetrical and Gynecological Survey*, by whose kind permission this abstract is reproduced here, remarks in one of his admirable commentaries: 'These cases of "suspended animation"... remind us that pronouncement of death in a newborn infant must be 100% certain.']

Convulsions in Childhood: I. Incidence and Classification

H. SCHONENBERG and W. E. GÖPEL. *Arztliche Wochenscrift*, 1958, **13**, 949–954.

In recent years the problem of convulsions in childhood has come more into the foreground, owing to the developments in electroencephalography and new drugs, and the greater attention being paid to the social and psychological aspects of the problem. The proportion of children admitted to the Aachen Children's Clinic for convulsions ranges from 4% to 11%, and epilepsy is as common as manifest tuberculosis and more common than diabetes in children. Convulsions in childhood can be divided into occasional spasms and chronic recurring convulsions or epilepsy. Both groups are further subdivided and discussed in detail.

(*Excerpta Medica* Abstract)

Van Creveld (*Amsterdam*)

Convulsions in Childhood: II. Treatment

H. SCHONENBURG and W. E. GÖPEL. *Arztliche Wochenschrift*, 1958, **13**, 973–977.

The aim of therapy is complete control of the seizures, thus avoiding intellectual and emotional damage. Each drug should be tried alone and in combination with others. Sudden discontinuation of a drug should be avoided. Bromine is no longer used. Barbiturates and the hydantoins (diphenylhydantoin, mesantoin) are effective in grand mal, oxazolines (tridione, paradione) especially in petit mal. Phenurone (a urea derivative) is effective in grand mal and petit mal but is too toxic to use in children. Pyrimidine derivatives (myelepsin) are effective in grand mal, psychomotor seizures and possibly in jacksonian epilepsy. Dietary therapy (low salt and water and ketogenic diets) is too troublesome and expensive to be used in children but is useful during the start of therapy. Psychotherapy for both children and parents is of great value, and it should include a clear and honest explanation to both of the nature of the illness, its treatment, and its possible course. In this way treatment is more likely to be accepted, and misconceptions can more easily be corrected. Overprotection and overindulgence should be avoided, and the child should be treated at home or in ordinary schools as a normal youngster. The term 'epilepsy' is best avoided, especially to the child, because of its prejudicial connotations. Surgery should be considered only if a focus is found, if medical treatment has proved ineffective, and if emotional and intellectual disturbances are increasing.

(*Excerpta Medica* Abstract)

Glaser (*Baltimore*)

Cerebral Palsy in Manitoba

J. K. MARTIN. *Canadian Medical Association Journal*, 1960, **82**, 411–417.

The data for this article are drawn from an analysis of the case-notes of 370 children with cerebral palsy. The great majority have been treated by the Society for Crippled Children and Adults of Manitoba. The greatest incidence of the disease is probably among those between 5 and 9 years old; it amounts to 1.574 per 1,000 (in some countries the frequency is expressed per 100,000 of the population, whence arise differences in quoted figures). Classification of cases by diagnosis showed that 73.6% were spastics and, in this group, the majority were right hemiplegias.

In research on the aetiology the author has confined himself to single factors, although in a number of cases there seems to have been more than one causal factor. Prematurity has been the abnormality most frequently encountered in the course of these investigations. Among acquired lesions, encephalitis (especially western equine encephalomyelitis), marked pyrexia, cranial injury and meningitis have been implicated in most cases. Anoxia was considered a factor if the history of birth

included cyanosis, prolonged labour, foetal distress or other causes suggestive of oxygen lack. In 22 cases (5.9%) Rh incompatibility was the only aetiological factor that could be isolated. The term 'familial' was used to characterise the disease when more than one child in the same family was affected. However, 10 children from five families were placed in other aetiological groups, for reasons given. Among the 216 cases of cerebral palsy considered congenital, 19 pairs of twins occurred. This frequency of 8.7% is nearly eight times the normal incidence of twins in the population; 25 patients had associated abnormalities.

The intelligence quotient of 33% of the children was less than 70, and 25.8% had an I.Q. between 70 and 89, with 41.2% ranging from 90 upwards. About 40% of the children could attend the ordinary schools, while about 30% needed special educational facilities. As far as intensive therapy was concerned, about 35% needed it and an equal proportion required little or no treatment at all. It would seem that 15% should be placed in special schools for backward children, while 15% may need institutional care at some time in their lives. The most optimistic experts in this field believe that not more than 60% of these infants would eventually be able entirely to fend for themselves.

(Author's summary, translated from the French.)

NOTE: *Dr. Cyrille Koupernik, of Paris, informs us that the paper abstracted on pages* 40–41 *of* '*Bulletin*' *No.* 8 *was written by him in French and translated into Russian by the Korsakow journal. It was an original paper never published in France, and the abstract is absolutely faithful. We apologise if anyone was confused by the Russian version of the author's name* ('*S. Kupernik*').—EDITOR, '*Cerebral Palsy Bulletin*'.

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